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Cardiology in the Young

WORLD CONGRESS OF PEDIATRIC CARDIOLOGY AND CARDIAC SURGERY

7th World Congress of Pediatric Cardiology & Cardiac Surgery Abstracts

ORAL ABSTRACTS

ADMINISTRATIVE

O1289 - IMPROVING TIMELY DISCHARGE BY USING GOAL ORIENTED ROUNDING AND TEAM BASED COMMUNICATION

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Background: Discharge planning for complex congenital heart patients is challenging for the multi-disciplinary team, especially in a cardiac critical care environment. Delays in discharge are often attributed to poor team planning and communication. Multi-disciplinary collaboration with effective communication ensures a seamless and timely discharge.
Methods: Members of the multi-disciplinary team collaborated to establish a rounding process in the intensive care unit to have patients discharged by noon. The rounding process, implemented in October 2015, had pre-defined goals and involved all members of the team: nursing, physicians, case management and allied health professionals. Patient status, eligibility for discharge and anticipated needs were discussed and planned accordingly by appropriate team members. Rounds were audited for goal completion and efficiency and patient discharge times were tracked.
Results: We identified four crucial multidisciplinary rounds: (1) Operational rounds which are brief overview rounds conducted in a conference room, establishing key goals for the day including preparing for the following days discharges, (2) Work rounds – bedside rounds, (3) 4 pm hand-off rounds – review of key goals days, and reviewing barriers to discharge and (4) Late evening rounds with bedside nurse to make sure goals are on track. Surveys were conducted at 1 month and 3 months after process education and implementation was completed. Responses were from all disciplines. There was an overall increase in process understanding, overall efficiency ($p < 0.1$). Prior to implementation of process, 25% patients were discharged prior to noon. From October 2015-September 2016, 40% of patients were discharged before noon.
Conclusion: Establishing a process for goal-oriented, multi-disciplinary rounding improves team communication and coordination of care in preparation for discharge. Ensuring timely discharge improves patient satisfaction and patient flow within the cardiac critical care unit.

O1660 - A CLINICAL EFFECTIVENESS PROGRAM LEVERAGING AGGREGATE PATIENT DATA AT THE POINT OF CARE IMPROVES VALUE IN PATIENTS UNDERGOING CONGENITAL HEART SURGERY

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Background: Healthcare in the United States is increasingly losing value. Decreasing variation is central to reducing waste and cost. The rapid adoption of information technology is regarded as an important means to promote high value care. We describe a clinical effectiveness program leveraging personalized comparative effectiveness information at the point of care to provide target hospital goals for patients following congenital heart surgery.
Methods: Using an observational pre-post-intervention design, patients undergoing one of the 10 core congenital heart surgeries as defined by the Society of Thoracic Surgeons were included. The setting was a tertiary university-affiliated academic children's hospital between September 6, 2016 and December 19, 2016. Personalized comparative cohorts encompassing 2 years of pre-intervention data were constructed utilizing the electronic health record (EHR). Based on the aggregate data, target hospitalization goals (e.g., target extubation time) were tailored for every patient and made visible peri-operatively for healthcare providers in real-time. Outcome metrics included intensive care unit (ICU), total post-operative length of stay (LOS) and associated cost.
Results: A total of 51 patients were enrolled with 47 (92%) completing the program. Other than the Fontan operation, all surgeries experienced a reduction in LOS. We found an aggregate reduction in LOS and variance for ICU (median 3 [IQR 3-4] vs 5 [IQR 3-7] days, $p < 0.001$; mean $3.6 + 1.9$ vs $6.5 + 6.5$ days, $p = 0.003$), and total postoperative LOS (median 6[IQR 5,8] vs 8[IQR 6,12] days, $p < 0.001$; mean $6.5 + 2.3$ vs $10.9 + 9.9$ days, $p = 0.003$), compared with the pre-intervention period. Mortality, reintubation, ICU and hospital readmission rates were unchanged. The annualized cost savings is estimated to be approximately $2.5 million.

Conclusions: Implementation of a clinical effectiveness program that leverages EHR data for real-time comparative effectiveness information decreased variation and safely reduced length of stay and associated cost.

O1973 - HOW HARD DO CARDIOLOGISTS WORK THE PATIENT ENCOUNTER INDEX (PEI) A NOVEL METHOD TO ASSESS THE CLINICAL WORKLOAD OF A PAEDIATRIC CARDIOLOGY SERVICE

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Background: Correctly assessing the workload of a clinical service may be difficult, particularly for paediatric cardiac services with many complex patients - when a simple count of patient numbers will be misrepresentative. We conceived a novel patient encounter index (PEI), which simply indexes the total daily encounter time as a ratio of a contractual 8-hour working day. A patient encounter is defined as any activity that may directly influence the clinical outcome of a specific patient.
Methods: Nine medical students observed all the daily clinical duties of four full-time cardiology clinicians over a 2-week period. The duration of each encounter and its nature (clinical, analytical, procedural, or discussive) were recorded. Encounters were recorded on pre-designed time sheets, transferred to Excel spread sheets and analysed using simple summary statistics.
Results: A total of 2014 encounters were recorded over 10 working days. On average, clinicians were involved in 45.3 (45.2-58.5) encounters per day, involving 30.9 (17.7-56.2) patients (some patients were encountered more than once per day). The four clinicians' PEIs ranged from 0.74 to 0.96. All encounters were timed from onset to completion; encounters required an average of 6.6 hours of dedicated time per day. The context and implications of these and other data (e.g. encounter intensity) will be presented.
Conclusions: Clinical duties consumed 74-96% of the working day of the clinicians, leaving little time for other essential duties, e.g. teaching and administration. When one clinician is away on leave, there is an 8% increase of each remaining clinician's clinical workload, resulting in PEIs of 0.82-1.04. These data were included into a motivation to increase clinical staffing of our cardiac services, with success. The PEI is a robust and repeatable tool to objectively assess clinical workload of busy clinicians, and may be effectively implemented to influence decisions in service management.

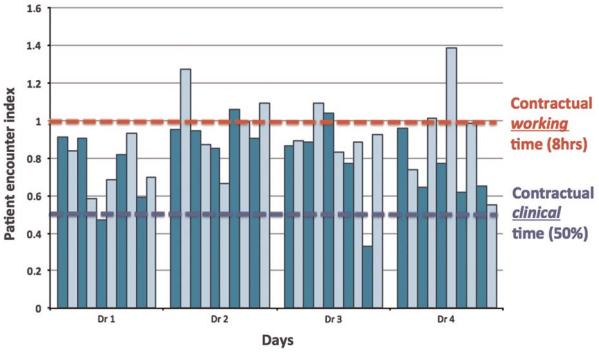

Figure.
Patient encounter index per doctor per day.

O2299 - PERFORMANCE OF PEDIATRIC CARDIAC SURGERY CENTERS A NATIONAL EFFORT TO REDUCE THE GAPS OF BURDEN OF CONGENITAL HEART DISEASE IN COLOMBIA

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Background: In Colombia about 4.800 new pediatric cardiac surgeries should be performed per year. Knowledge of the characteristics and results of Pediatric Cardiac Surgery Centers in Colombia allows us to determine: The real number of patients treated and the capacity of our country to meet the needs of patients with congenital heart disease to promote new strategies in this field. The aim of this study was to describe the operational capacity of Pediatric Cardiac Surgery Centers in Colombia for 2015.
Methods: A blind online survey was sent to the centers previous a verbal agreement to the willing of participate. The survey had 25 questions related to 4 domains: volume of procedures, services offered, surgical risk evaluation and databases. A descriptive analysis was done in STATA 14.
Results: 13 Pediatric Cardiac Surgery Centers were identified in Colombia and all answered the survey. 38,5% (5/13) are in Bogota, follow by Bucaramanga and Cali with 2 centers each one of them. All centers had pediatric cath lab, int 2015 a total of 2406 cardiac surgeries were done, a mean of 55.6% of all surgeries were on pump, and centers reported 157 adult congenital heart surgeries. The 46% of the centers had only one dedicated surgeon for these surgeries. In 53.8% (7/13) of the centers, all RACHS-1 category procedures are performed. Same proportion of centers had available ventricular assistance devices. The 61.5% (8/13) centers had own database, only 2 centers are reporting to international audit database.
Conclusions: In order to promote changes in the care of our patients we need information. In developing countries, this is one of the most difficult problems, however with the commitment of the surgeons that lead the institutions in Colombia, we could collect this very important information.

ADULT CONGENITAL HEART DISEASE

O1064 - FACTORS ASSOCIATED WITH LONG TERM MORTALITY AFTER FONTAN PROCEDURES. RISK SCORE CREATIOIN BASED ON SYTEMATIC REVIEW OF 6707 PATIENTS FROM 28 STUDIES

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Background: Despite an aging Fontan population, data on late outcomes is still scarce. Reported outcome measures and determinants vary greatly between studies making comprehensive appraisal of mortality hazard challenging.
Methods: We conducted a systematic review to evaluate causes and factors associated with late mortality in patients with Fontan circulation. Late mortality was defined as mortality beyond the first postoperative year. Studies were included if they had >90 patients or >20 late mortalities and/or transplants. Studies with overlapping patients were rationalized to include only the most recent studies to avoid duplication. The pooled hazard ratios were used to create the initial components and weighting of a clinical score for Fontan patients.
Results: From 28 studies, a total of 6707 patients with an average follow-up time of 8.23 ± 5.42years were identified. There were 1000 deaths. Causes of late death were reported in 697 cases. The five most common causes were heart/Fontan failure (22%), arrhythmia (16%), respiratory failure (15%), renal disease (12%), and thrombosis/bleeding (10%). Factors associated with late mortality were evaluated and classified into 9 categories and their relative hazards were used to derive the initial components of a weighted, practical and clinically based Fontan risk score (ranging from 0-100). Nine risk factors including anatomic risk factors, elevated preoperative pulmonary artery pressure, age >7 at the time of the operation, atriopulmonary Fontan, heart failure symptoms, arrhythmia, moderate/ severe ventricular dysfunction or atrioventricular valve regurgitation, protein losing enteropathy and end organ disease (cirrhosis or renal insufficiency) were identified as the most significant.
Conclusion: Factors associated with late mortality after the Fontan operation are summarized in this study, and the components of a new mortality risk score, that is weighted, and potentially usable in an outpatient setting are presented. Prospective validation and refinement of this risk score will be undertaken.

O1148 - PHYSICAL ACTIVITY ASSESSMENT IN PATIENTS WITH CONGENITAL HEART DISEASE AND ASSOCIATIONS TO EXERCISE CAPACITY AND HEALTH RELATED QUALITY OF LIFE

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Objective: Improved survival leads to growing importance of functional outcome measures in patients with congenital heart disease (CHD). This study applied the International Physical Activity Questionnaire (IPAQ) to assess self-reported physical activity in patients with CHD and their association to exercise capacity and health-related quality of life (HrQoL).
Patients and Methods: Prior to cardiopulmonary exercise testing (CPET), 786 consecutive patients (335 female, 31.1 ± 11.6 years) with CHD filled in the short form of the IPAQ and the HrQoL questionnaire SF-36.
Results: In total, 393 (50.0%) patients reported health enhancing physical activity (HEPA), 237 (30.2%) minimal activity and 156 (19.8%) inactivity. In comparison to the HEPA group the inactive group had significant lower peak oxygen uptake (74.2% ± 20.7% vs. 86.8% ± 22.3%; $p < .001$) as well as lower physical (91.0% ± 16.9% vs. 97.4% ± 13.6%; $p < .001$) and mental (97.1% ± 22.2% vs. 104.1% ± 15.6%; $p < .001$) HrQoL. Independent for severity class, surgery, age, beta blocker, pacemaker and oxygen saturation, the HEPA group showed in comparison to the inactive group significant less probability for impairments in mental (OR: 0.21 CI: 0.10–0.46; $p < .001$) and physical (OR: 0.49 CI: 0.25–0.97; $p = .039$) HrQoL and exercise capacity (OR: 0.38 CI: 0.24–0.61; $p < .001$).
Conclusions: Categorization of CHD patients with the IPAQ provides clinical information since HEPA patients have a less probability for impaired HrQoL and reduced exercise capacity. Nevertheless, the IPAQ cannot substitute an accelerometer based assessment for physical activity, nor a CPET for exercise capacity.

O1309 - CONGENITAL HEART DISEASE AND RISK OF INFECTIONS IN THE CENTRAL NERVOUS SYSTEM

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Background: Central nervous system (CNS) infections are associated with substantial morbidity and mortality. Neurologic complications are prevalent in the congenital heart disease (CHD) population, however the risk for CNS infection is unknown. We aimed to compare the risk of CNS infection in the CHD population with the general population.
Materials and Methods: In this population-based cohort study, we used medical registries covering all Danish hospitals to identify subjects with CHD diagnosed before the age of 5 years, born between 1958 and 2000. Subjects were followed until first-time hospital-diagnosis of any CNS infection, death, emigration, or end of study (December 2012). For each CHD subject, we identified 10 individuals from the general population utilizing the Danish Civil Registration System, matched on sex and birth year. We computed cumulative incidences with death as a competing risk and hazard ratios (HRs) with corresponding 95% confidence intervals (CIs) adjusted for birth year and sex.
Results: The CHD cohort consisted of 10,046 individuals (51% male). Among subjects with CHD, cumulative incidence of any CNS infection by age 30 years was 1.0% and 2.5% for endocarditis, a known risk factor of CNS infection. The overall HR of CNS infection was 2.7 (95% CI: 2.1-3.4) among CHD subjects compared to the general population cohort. The HR for mild CHD was 2.1 (95% CI: 1.4-3.3), moderate CHD 1.5 (95% CI: 0.8-2.6), and severe CHD, including univentricular physiology, 3.9 (95% CI: 2.7-5.6). Lesions with a cyanotic history demonstrated a HR of 4.2 (95% CI: 2.5-7.2). Among CHD subjects without documented record of endocarditis, the risk of CNS infection remained elevated (HR: 2.6; 95% CI: 2.1-3.3). HRs specific to meningitis and cerebral abscesses were 2.5 (95% CI: 1.8-3.5) and 5.0 (95% CI: 3.3-7.4), respectively.
Conclusions: Subjects with CHD experienced an increased risk of CNS infection compared with the general population.

O1339 - ABNORMAL PLASMA LIPID PROFILE IS ASSOCIATED WITH POOR PROGNOSIS IN FONTAN PATIENTS

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Background: Low plasma levels of total cholesterol (TC), low and high density lipoprotein cholesterols (LDL, HDL) characterize lipid profile in patients after Fontan operation. However, the clinical significance of this abnormality remains unknown.
Purpose: This study was conducted to clarify associations of the abnormal lipid profile with Fontan pathophysiology.
Method and Results: We measured plasma levels of those cholesterols and calculated arteriosclerosis index (AI) in 340 Fontan patients and 28 controls and compared the results with the clinical profiles, including hemodynamics and prognosis. Fontan patients showed lower TC, LDL, HDL ($p < 0.0001$) and similar AI when compared with those in controls. Male gender, low body mass index (BMI), presence of protein losing enteropathy (PLE), and high central venous pressure independently predicted lower LDL ($p < 0.05$-0.001), whereas PLE was the only independent predictor of lower HDL ($p = 0.03$). BMI, PLE, and HbA1c levels independently predicted higher AI ($p < 0.05$-0.0001). During the follow-up, 108 (22 deaths) clinical events occurred. Low HDL and High AI predicted morbidity ($p < 0.05$-0.0001). Furthermore, bivariate model with these variables with brain natriuretic peptide (BNP) or peak oxygen uptake (VO2) revealed that the prognostic value of high AI was independent of these two established prognostic variables ($p < 0.05$ for both models).
Conclusions: Fontan patients showed low TC, LDL, and HDL and high AI, especially low HDL and high AI in PLE patients. Increased AI and low HDL had significant prognostic power regardless of the hemodynamics.

O1351 - SURGICAL STRATEGIES OF PULMONARY VALVE REPLACEMENT AS RE DO PROCEDURE IN PATIENTS WITH CONGENITAL HEART DISEASE

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Objectives: Many patients with congenital heart disease (CHD) undergo surgical pulmonary valve replacement (S-PVR) due to pulmonary regurgitation after right ventricular outflow tract (RVOT) repair. We compared beating and arrested heart techniques for PVR.
Methods: Data of 57 patients who underwent S-PVR between 2012 and 2016 was analyzed retrospectively.
Results: S-PVR was performed at mean age of 23.6 ± 18.6 years, 21.2 ± 13.5 years after initial surgery for CHD. Indications for re-operation were pulmonary insufficiency in patients with TOF ($n = 39$), congenital pulmonary stenosis/atresia ($n = 11$), TGA ($n = 4$), truncus arteriosus ($n = 1$), previous Ross procedure ($n = 1$) and RVOT reconstruction ($n = 1$). Three different bioprostheses were implanted: Carpentier-Edwards Perimount ($n = 32$), St. Jude Trifecta™ ($n = 23$) and BioIntegral Surgical Bioconduit™ ($n = 1$) measuring 19-27mm. Tricuspid valve annuloplasty was necessary in 11 patients (20%). Surgery was either performed with beating ($n = 28$, Group_A) or arrested heart ($n = 29$, Group_B) technique. Especially in Group_B right ventricular (RV) function was impaired (69%). Cardiopulmonary bypass (CPB) duration differed significantly between the groups who exclusively received S-PVR with aortic cross-clamping (134 ± 75 minutes) and with beating heart (76 ± 38 minutes, $p < 0.05$). 68% of Group_A patients received St. Jude Trifecta™ bioprosthesis and 83% of Group_B patients Carpentier-Edwards Perimount bioprostheses. Median time to extubation was shorter in Group_A (6 hours), compared to Group_B (12 hours, $p = 0.22$) while median ICU-stay was comparable in both groups (Group_A: 2 days; Group_B: 3 days). Patients were discharged after an average in-hospital stay of 11 days (Group_A) and 15 days (Group_B), respectively ($p < 0.05$). 30-day-mortality was the same in both groups (Group_A: 3.6%; Group_B: 3.4%).
Conclusion: S-PVR is common in patients with CHD. Beating heart technique has favorable effects on duration of intubation, ICU and in-hospital stay. Extrathoracic cannulation before sternotomy was used in Group_B and thus CPB duration was significantly longer. Beating heart technique should be used whenever possible, despite impaired RV function.

O1393 - SERUM CARBOHYDRATE ANTIGEN 125 LEVEL PREDICTS FUTURE CARDIOVASCULAR EVENTS IN PATIENTS WITH FONTAN CIRCULATION

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Background: Plasma carbohydrate antigen-125 (CA-125) level has emerged as a biomarker and predictive factor for acute heart failure with normal-structure heart disease. However, the clinical significance of plasma CA-125 level in Fontan circulation remains unclear.
Materials and Methods: We prospectively enrolled 73 consecutive patients older than 15 years with Fontan circulation who were hospitalized between February 2015 and December 2016. Plasma CA-125 level and other biochemical markers were routinely tested within 24 hours of hospitalization. Patients were followed up until the following cardiovascular events occurred: acute or exacerbation of chronic heart failure, clinically significant arrhythmia, or thromboembolism.
Results: The median serum CA-125 level was 25 U/mL (range: 6-678), and was significantly higher in patients with higher New York Heart Association class (NYHA I/II: 21.5 vs. NYHA III: 131, $p = 0.0008$), and those with pretibial edema, pleural effusion, and ascites (160.5 vs. 19.5, $p < 0.0001$/141 vs. 21, $p = 0.0002$/85.5 vs. 19, $p < 0.0001$, respectively). During the follow-up period (9.4 ± 5.0 months), 18 cardiovascular events occurred (10 acute exacerbations of heart failure, 7 arrhythmias, and 1 cerebral thromboembolism). Univariate Cox regression analysis identified 8 clinical predictors ($p < 0.005$): history of protein-losing enteropathy (hazard ratio [HR]: 16.9), CA-125 > 36 U/mL (HR 11.5), aldosterone >298 pg/mL (HR 5.09), dopamine >7 ng/mL (HR 6.11), albumin <3.7 g/dL (HR 7.54), troponin T > 0.006 ng/mL (HR 7.97), sodium <139 mEq/L (HR 4.79), and administration of loop diuretics (HR 4.28). The clinical cut-off points were determined from the receiver operating characteristic curve. Multivariate analysis showed that CA-125 > 36 U/mL ($p < 0.0001$) and aldosterone >298 pg/mL (p 0.025) were independent predictive factors.
Conclusion: In patients with Fontan circulation, the CA-125 level correlates with fluid retention and predicts future cardiovascular events.

O1502 - EXPECTATIONS FOR SURVIVAL AND COMPLICATIONS FOR FONTAN PATIENTS WHO TRANSITION TO ADULT CHD CARE

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Background: Long term outcomes of Fontan patients who survive to age 16 years have not been well characterised. The Australia and New Zealand Fontan Registry (ANZFR) follows all Fontan patients in ANZ from childhood and provides a unique opportunity to understand survival and complication rates in Fontan patients who reach 16 years of age and beyond.
Methods and Results: Follow up of all 683 survivors aged 16 years and older who underwent a Fontan procedure in ANZ was analysed. Mortality status was confirmed from a National Death Registry. Mean age of the cohort was 26 8 years; There were 201 atriopulmonary connections (APC), 249 lateral tunnels (LT) and 233 extracardiac conduits (ECC). For those surviving to at least age 16 years, survival at 20 years of age was 96% (95% CI, 95%-98%), 90% (95% CI 87%-93%) at 30 years and 81% (95% CI 76%-87%) at 40 years of age. Survival was significantly worse for the APC patients compared to either type of TCPC; at 30 years of age 87% for APC versus 93% for TCPC; at 40 years, 78% for APC versus 86% for TCPC ($p < 0.01$). At 50 years of age, survival in the APC group was 61% (95% CI, 46-81%); insufficient TCPC patients had survived to that age to allow meaningful survival statistics. At latest follow up, only 53% were in NYHA Class I. After the age of 16 years, 136 (20%) had experienced at least one new arrhythmia, 41 (6%) required a permanent pacemaker, 10 (2%) experienced protein losing enteropathy, 45(7%) had a thromboembolic event and 131 (21%) required a surgical re-intervention.
Conclusions: Increasing numbers of patients with Fontan procedures are entering adulthood. This comprehensively followed shows that a variety of morbid complications is common in Fontan adults, and that there is progressive attrition, particularly in the APC subjects.

O1543 - VORTEX FLOW IN THE RIGHT ATRIUM SURROGATES SUPRAVENTRICULAR ARRHYTHMIA AND THROMBUS AFTER ATRIO PULMONARY CONNECTION TYPE FONTAN OPERATION A NOVEL METHOD OF VORTEX FLOW ANALYSIS USING CONVENTIONAL CINE MAGNETIC RESONANCE IMAGING

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Background: We developed a novel imaging technique, named vortex flow (VF) mapping, which demonstrates vortex flow visually on conventional two-dimensional (2D) cine MRI. We assessed circumferential VF patterns and the influences on RA thrombus and supraventricular tachycardia (SVT) in AP connection type Fontan circulation.
Method: Retrospectively, we enrolled 27 consecutive patients (25.1 +/- 9.2 years) and 7 age-matched controls who underwent cardiac MRI. Conventional cine images acquired on 1.5-tesla scanner were scanned for axial and coronal cross-section of the RA. We developed a "vortex flow mapping" which can demonstrate the ratio of the circumferential voxel movement at each phase to the total movement throughout a cardiac cycle towards the center of the RA. The maximum ratio was used as a magnitude of vortex flow (MVF %) in the RA cine imaging. We also measured a percentage of strong and weak VF areas (VFA %).
Results: 15 patients had cardiovascular complications (Group A) and 12 patients did not (Group B). On a transaxial image, strong VFA % in Group A was significantly smaller than that in Group B and controls. On a coronal view, strong VFA % was also smaller, and weak VFA % was larger in Group A compared to them in Group B and controls ($P < 0.05$, and $P < 0.05$). Maximum MVF % in Group A was significantly smaller than other groups ($P < 0.001$). On univariate logistic analyses, weak VFA % on a coronal image, and serum total bilirubin level were important factors of cardiovascular complications (Odds ratio 1.14 and 66.1. 95%CI 1.004-1.30 and 1.59-2755.6. P value <0.05 and <0.05, respectively).
Conclusions: Circumferentially weak VFA % on a coronal image could be one of the surrogate makers of SVT and thrombus in AP connection type Fontan circulation. This VF assessment is simple and clinically useful to detect blood stagnation.

O1571 - MYOCARDIAL PERFUSION ABNORMALITY BY SPECT IMAGING CORRELATES WITH VENTRICULAR MECHANO ELECTRICAL PROPERTY IN ADULT WITH CONGENITAL HEART DISEASE

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Background: Myocardial perfusion imaging (MPI) by single-photo emission computed tomography (SPECT) is an established modality to cardiac performance related with myocardial ischemia and/or infarction. However, the clinical role of MPI remains unclear in patients with adult congenital heart disease (ACHD).
Purpose: This study was to clarify the clinical significance of MPI by SPECT for evaluating the mechanical function and electrical property of the systemic ventricle (SV) in ACHD patients.
Methods and Results: Between August 2014 and February 2016, Resting SPECT MPI was performed in 95 ACHD patients (age: 31 ± 10 years, biventricular physiology in 69 and Fontan physiology in 26). We developed MPI defect score (%D: %) as an index of severity of the SV myocardial perfusion abnormality and compared the index with plasma levels of high-sensitive troponin T (hsTnT), brain natriuretic peptide (BNP), end-diastolic volume index and ejection fraction of the SV (EF), QRS duration, and peak oxygen uptake (ml/kg/min). In all ACHD patients, %D was 8.7 ± 8.4, ranged from 0 to 33.8. %D was independently correlated with positively with QRS duration ($p < 0.05$) and negatively with EF ($p < 0.001$) and inversely correlated with PVO2 ($p < 0.05$), whereas %D was not correlated with hsTnT or BNP. These associations remained significant in both ACHD patients with biventricular ($p < 0.05$-0.001) and those with Fontan physiologies ($p < 0.05$), except for an association of %D with EF in Fontan physiology.
Conclusion: SV myocardial perfusion abnormality was common in ACHD patients regardless of the hemodynamic pathophysiology. SPECT-PMI has a significant role for providing us a vital information on SV myocardial perfusion abnormality which is closely associated with SV mechano-electrial property in ACHD patients.

O1578 - ATTENTION DEFICIT HYPERACTIVITY DISORDER IN LARGE ASIA CONGENITAL HEART DISEASE COHORT

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Background: Incidence of late neurodevelopmental disorders as attention deficit hyperactivity disorder (ADHD) was reported to be higher in certain congenital heart disease (CHD) diseases. However, no large scales studies and no relevant studies in Asian population were reported before. We try to define the incidence and risk factors of ADHD in CHD patients.
Methods: From Jun 2015 to May 2016, all CHD patients, aged between 6-15 years, who visited the out-patient clinics in National Taiwan University Hospital were enrolled. We used Swanson, Nolan, and Pelham rating scale (SNAP-IV) Chinese version, which was validated effective, to analyze the three dimensions (inattention, hyperactivity, impulsivity) of ADHD and correlated with previous medical records.
Results: Totally 695 CHD patients (M/F: 372/323) were enrolled in our study with mean age 10.2 ± 2.43 years old. Cyanotic heart disease accounted for 217 patients (31.3%), inclusive of single ventricle 38 patients (5.5%). The incidence of inattention, hyperactivity, and impulsivity were 9.8%, 8.6% and 7.6%, respectively, which showed no significant difference with normal age-matched population. However, the incidence of inattention dimension in cyanotic CHD is 13.8%, which was significant higher than acyanotic CHD (7.9%, $p = 0.019$), especially in single ventricle group (incidence 23.7%, $p = 0.008$). For the hyperactivity and impulsivity dimension, the incidence showed no difference. The risk factors of inattention dimension of ADHD includes post-operative seizure, single ventricle, prolonged cyanosis, multiple operations and cardiopulmonary bypass, and postoperative extracorporeal membrane oxygenation (ECMO) support. Using multivariate regression analysis, post-operative seizure, single ventricle disease and postoperative ECMO support were important risk factors of late inattention disorder.
Conclusion: The overall incidence of ADHD is similar between CHD patients and normal population, but the incidence of inattention dimension is higher in single ventricle CHD and cyanotic CHD. The important risk factors of inattention disorder include post-operative seizure, single ventricle CHD and post-operative ECMO support.

O1582 - FAMILIAL CO OCCURRENCE OF CONGENITAL HEART DEFECTS FOLLOWS DISTINCT PATTERNS

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Aims: Congenital heart defects (CHD) affect almost 1% of all live born children and the number of adults with CHD is increasing. In families where CHD has occurred previously, estimates of recurrence risk, and the type of recurring malformation are important for counseling and clinical decision making, but the recurrence patterns in families are poorly understood. We aimed to determine recurrence patterns, by investigating the co-occurrences of congenital heart defects in 1,163 families with 3,080 individuals who had a clinically confirmed diagnosis.
Methods and Results: We calculated rates of concordance and discordance for 41 specific types of malformations, observing a high variability in the rates of concordance and discordance. By calculating Odds Ratios for each of 1,640 pairs of discordant lesions observed between affected family members, we were able to identify 178 pairs of malformations that co-occurred significantly more or less often than expected in families. The data show that distinct groups of cardiac malformations co-occur in families, suggesting influence from underlying developmental mechanisms. Analysis of human and mouse susceptibility genes showed that they were shared in 19% and 20% of pairs of co-occurring discordant malformations, respectively, but none of malformations that rarely co-occur, suggesting that a significant proportion of co-occurring lesions in families is caused by overlapping susceptibility genes.
Conclusion: Familial congenital heart defects follow specific patterns of recurrence, suggesting a strong influence from genetically regulated developmental mechanisms. Co-occurrence of malformations in families is caused by shared susceptibility genes.

O1595 - ARRHYTHMIAS AND SPECIAL ELECTROPHYSIOLOGICAL CHARACTERISTICS IN CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

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Introduction: Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart defect. It may associate with complete atrioventricular block (cAVB) and a variety of arrhythmias but the exact mechanisms are still not well-elucidated.
Methods: We reviewed medical records of patients diagnosed with ccTGA and follow-up in our institution from 1998 to 2016. Supraventricular tachycardia (SVT) in our study is defined as paroxysmal SVT (atrioventricular reentrant tachycardia, atrioventricular nodal reentrant tachycardia AVNRT, and twin AVNRT) and atrial arrhythmias.
Results: Ninety-two patients (37 female; 55 male) were enrolled in our study. The 40-year transplant free survival was 78.7% after 1922 patient-years of follow-up: mortality rate was 13% and 4.3% of patients received heart transplant. The 20-year and 40-year SVT free survival rate were 70.4% and 50.5%, respectively. The 20-year and 40-year cAVB free survival rates were 92.2% and 76.8%, respectively. For 31 patients (33.7%) with SVT, paroxysmal SVT accounts for 15 (16.3%) and atrial arrhythmia accounts for 18 (19.6%) with 2 (2.2%) had concomitant both kinds of SVT. Seven patients (7.6%) developed cAVB during follow-up. Neither cardiac operation history nor anatomical features were significantly associated with development of SVT or cAVB. Twenty-two patients received a total of 31 electrophysiological studies for SVT. The mechanisms of SVT include atrial arrhythmia in

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6 (33.3%) and paroxysmal SVT in 10 (45.5%). The mechanisms of paroxysmal SVT include concealed right-side accessory pathways in 3 (25%), manifest left-side Kent pathways in 2 (16.7%), twin AVNRT in 3 (25%), and AVNRT in 4 (33.3%). Catheter ablation successfully eliminated these paroxysmal SVT in 6 (60%) patients.
Conclusions: Arrhythmia burden, including paroxysmal SVT, atrial arrhythmia, and cAVB, in patients with ccTGA is high and increases over time. The mechanisms of SVT are complicated especially for paroxysmal SVT and can be controlled in some by catheter ablation.

O1596 - THE LYMPHATIC MORPHOLOGY AND FUNCTION IN FONTAN OPERATED PATIENTS

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Background: The lymphatics regulate the interstitial fluid by removing excessive fluid. It represents an extremely important step in the prevention of edema. The Fontan procedure has revolutionized the treatment of univentricular hearts. However, it is associated with severe complications such as protein-losing enteropathy (PLE) and peripheral edema that may involve the lymphatic circulation.
Hypothesis: Patients with a univentricular circulation have a reduced functionality of the lymphatic vasculature, which predisposes them to developing complications such as edema and PLE.
Material and Methods: The functional state of lymphatics is investigated using near infrared fluorescence imaging, NIRF. The anatomy is described using non-contrast MRI and the capillary filtration rate is measured using plethysmography. The study population is patients with Fontan circulation operated at Aarhus University hospital. Exclusion criteria is BMI > 30 and age (years) < 18. The Fontan group will be compared with an age, gender and weight matched control group of healthy volunteers.

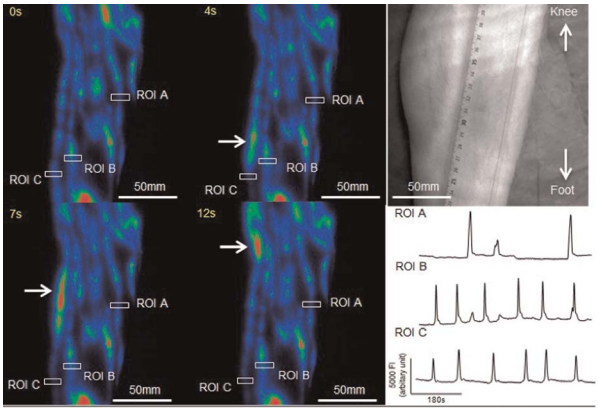

Figure 1.
Near infrared fluorescence imaging with indocyanine green enables lymphatic contractile function to be measured in vivo. *The image series depicts a network of lymphatic vessels on the lower leg. An example of a packet of lymph moving cranially is marked with a white arrow. Top right is a photograph of the same area. Bottom right shows an intensity plot over a longer time course for ROI A, B and C. Pulsatile changes in fluorescence intensity (FI) can be seen, reflecting lymphatic contractions. Images are pseudocoloured rainbowscale with blue and red indicating the least respectively most intense signal.*

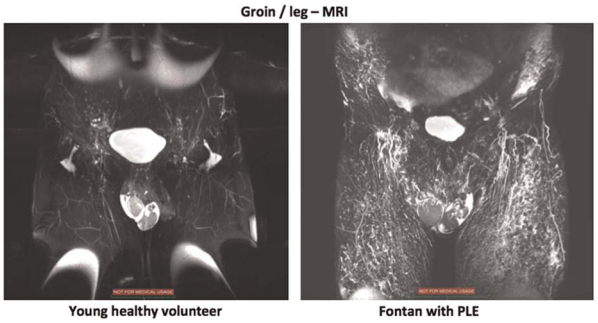

Figure 2.
MRI of lymphatic vessels in the groin area of a healthy control (left) and a Fotan patient (right). The two images depicts the lymphatic vessels in the groin area visualized by MRI. It is clear that the Fontan patient has a massive amount of lymphatic vessels and a abnormal collaterization compared to the healthy volunteer.

Results: The results (Fontan n = 7, Control n = 7) show that Fontan patients with clinical edema have a vast abnormal network of lymphatic collaterization. We also find a dilated thoracic duct with an abnormal course compared to normal. Baseline values found in the control group using NIRF, show that the lymphatic vessels have a contraction frequency: 0.6(0.2) min-1, propulsion velocity: 1.8(0.2) cm/s and are able to generate a pumping pressure: 57(3.2) mmHg. These values correlates with values found in in-vitro experiments. In comparison lymphatic function in the Fontan group using NIRF, shows a contraction frequency: 0.9 (0.2) min-1, propulsion velocity: 2,2(0.4) cm/s and a pumping pressure: 53(3.7) mmHg. No significant difference is seen for these parameters between Fontan patients and controls, $p > 0.05$.
Conclusion: The results indicate that lymphatics in some Fontan patients are abnormal with respect to morphology while function seems not to be altered.

O1751 - MANAGEMENT ERRORS IN ADULTS WITH CONGENITAL HEART DISEASE – PREVALENCE SOURCES AND CONSEQUENCES

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Background: Improved survival has resulted in increasing number and complexity of adults with congenital heart disease (ACHD). Guidelines recommend specialised care but many are still not managed at dedicated ACHD centres.
Methods: We analysed referral sources and appropriateness of management for patients referred to our tertiary-based ACHD Centre over the past 3 years (30 ± 13 years, 44% male). Only new patients ≥16 years with CHD, seen by another cardiologist within 3 years, were included. We compared differences in care between paediatric/ACHD-trained versus general adult cardiology referrals, according to Compliance (C) or Non-Compliance (NC) with published guidelines. Patients referred directly when CHD was identified were classified as C. NC cases were graded according to adverse outcome or risk of adverse outcome; Catastrophic (death/near-death/permanent disability); Major (permanent loss of function/recommendation for unnecessary intervention/failure to

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recommend necessary intervention exposing patient to unnecessary risk of death); Moderate (less serious loss of function/risk) or minor (failure to meet guidelines concerning diagnosis/treatment).
Results: Of 229 patients included, 128 (56%) were general cardiology referrals (58% with complex CHD) and 101 (44%) were paediatric/ACHD referrals (67% with complex CHD). Forty-seven (37%) general cardiology referrals were NC; 18 (14%) had catastrophic or major issues (n = 2,16 respectively), 17 (13%) moderate and 12 (9%) minor. Twelve (12%) paediatric/ACHD referrals were NC but none were catastrophic, 2 were major, 1 moderate and 9 minor. Deviations from guidelines were far more common in referrals from general compared to CHD-trained ($p < 0.0001$) as were catastrophic/major complications ($p = 0.002$). All catastrophic and 56% of major issues occurred in complex CHD. The most common catastrophic/major issue was mismanaged severe pulmonary regurgitation.
Conclusions: Over half referrals to a specialised ACHD Centre were from general cardiologists, 37% were NC with guidelines, often with catastrophic or major consequences. Adequate access to specialised service provision for this growing population is essential.

O1944 - THE IMPACT OF POST OPERATIVE ACUTE KIDNEY INJURY FOLLOWING FONTAN CONVERSION SURGERY

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Ann & Robert H Lurie Children, Pediatric Cardiology, Chicago-United States[1]; Northwestern University Feinberg School of Medicine, Biostatistics, Chicago-United States[2]; Ann & Robert H Lurie Children, Cardiovascular-Thoracic Surgery, Chicago-United States[3]; Stanford University Medical Center, Pediatric Cardiology, Palo Alto-United States[4]; Ann & Robert H Lurie Children, Kidney Diseases, Chicago-United States[5]

Background: Fontan conversion (FC) is a useful option for "failing Fontan" physiology. Acute Kidney Injury (AKI) is associated with worse outcomes after cardiac surgery. We evaluated the incidence, predictors of, and prognostic value of AKI following FC.
Methods: This single-center retrospective cohort study included FC patients from 1994-2016. AKI was classified using KDIGO criteria: AKI 1 = 1.5-2, AKI 2 = 2-3, AKI 3 = > 3 fold increase in serum-creatinine (sCr) within 7-days post-op. Multivariable logistic regression was created to identify risk factors for AKI ≥ 2. Associations between AKI ≥ 2 and post-operative outcomes were evaluated using t-tests. Heart-transplant-free survival among AKI groups was compared using Kaplan-Meier curves and log-rank test.
Results: Included were 139 FC patients: mean age at FC 25 ± 9 years; 81% atrio-pulmonary; 86% left-ventricular morphology; median follow-up after FC 8 (0.02-21) years. Pre-op sCr was normal for all patients. After FC, 63 patients (45%) developed AKI, including AKI 1 in 37(27%), AKI 2 in 10(7%), and AKI 3 in 16 (11%). Renal function recovered (sCr <1.5 times compared to pre-op) by hospital discharge in 86%, 80%, and 19% of patients with AKI 1, 2, and 3, respectively. Independent risk factors for AKI ≥ 2 included higher number of prior sternotomies (OR = 1.97; 95% CI = 1.05-3.68); higher right atrial pressure (OR = 1.18; 95% CI = 1.02-1.37), and prior ablation procedure (OR = 3.45; 95% CI = 1.17-10.18). AKI ≥ 2 was associated with longer postoperative hospital length-of-stay [median 18(8-135) vs. 10(6-58) days; $p < 0.001$]; more chest tube days [9 (5-57) vs. 7(3-28) days; $p = 0.01$]; longer intubation time [median 2(1-117) vs.1(1-6) days; $p = 0.01$], and greater need for dialysis (31% vs. 0%; $p < 0.001$). Greater severity of AKI was associated with worse intermediate-term heart-transplant free survival (Figure 1) and overall survival (figure 2).
Conclusion: Nearly half of patients develop Acute Kidney Injury following Fontan Conversion. AKI ≥ 2 is associated with worse outcomes and reduced survival following FC surgery. Knowledge of AKI predictors may allow for improved FC risk stratification, patient selection, and perioperative management.

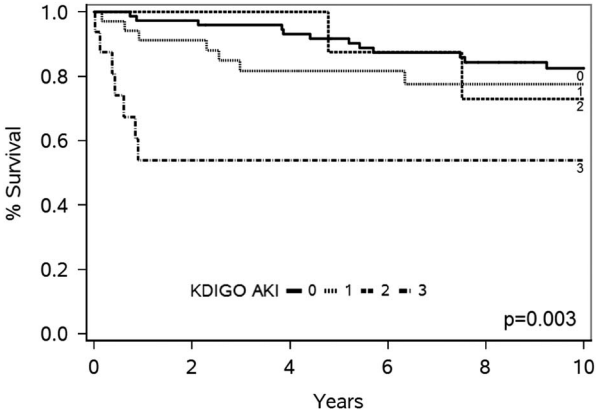

Figure 1.
Heart Transplant Free Survival by AKI Severity.

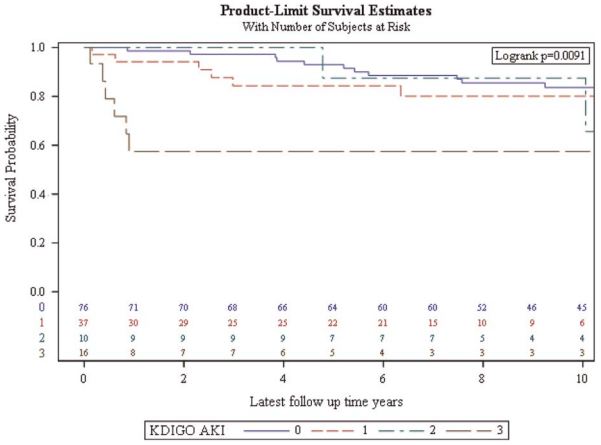

Figure 2.
Overall survival by AKI severity.

O1962 - SURGICAL OUTCOME FOR COMPLICATIONS AFTER REPAIR IN ADULT CONGENITAL HEART DISEASE

Kosuke Yoshizawa[1], Keiichi Fujiwara[1], Nobuhisa Ono[1], Hisanori Sakazaki[2]
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Background: Surgeries for adult congenital heart disease (ACHD) are mainly divided into two groups, repair surgeries in adulthood or surgeries for late complications after repair.
Objectives: To investigate our surgical outcome in ACHD patients with complications after repair (definitive repair or Fontan type operation).
Methods: ACHD patients (age ≥ 18) who underwent major cardiovascular operations between 1991 and 2016 were studied (n = 216, mean age: 35.6 years old, mean follow up: 8.2 years). We identified 82 surgical cases for complications after previous repair

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(Group1). Main surgical indications include RVOT related (n = 39, with Bentall's operation n = 3), TCPC conversion (n = 10), aneurysm after CoA repair (n = 4), and cardiac resynchronization therapy (CRT) (n = 11). Other 134 cases underwent the repair operation in adulthood (Group 2).
Results: Hospital deaths (within 30 days) were found in 3.7% (Group 1) and 2.3% (Group 2) respectively. Group1 had lower late survival rate than Group 2 (log-rank, p = 0.02) and preoperative NYHA class III or IV was detected as a risk factor for overall mortality (p = 0.01).
Conclusion: Our overall surgical outcome in ACHD patients was feasible, although surgeries for complications after repair are still complex. Surgeries of patients with preoperative severe heart failure are challenging.

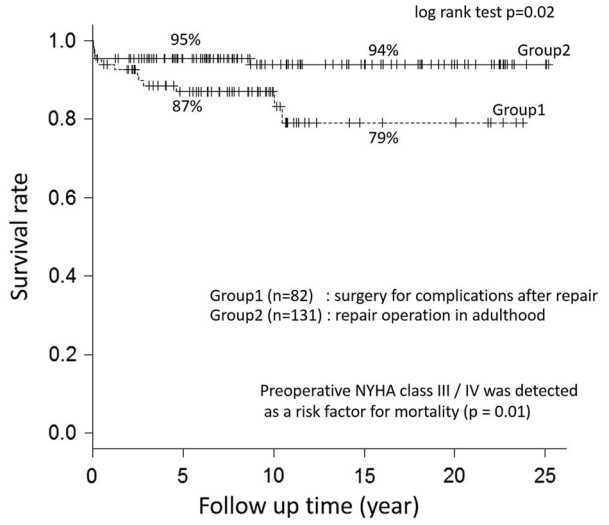

Figure.

O2043 - BURDEN OF AND RISK FACTORS FOR ACUTE KIDNEY INJURY IN ADULTS WITH CONGENITAL HEART DISEASE UNDERGOING CARDIAC SURGERY

Rohit Loomba[1], Christopher Learn[1], Lisa Dipietro[1], Stuart Goldstein[2], David Cooper[1]
Cincinnati Children's Hospital, Cardiology, Cincinnati-United States[1]; Cincinnati Children's Hospital, Renal, Cincinnati-United States[2]

Background: Postoperative acute kidney injury (AKI) in adults with congenital heart disease (ACHD) undergoing cardiac surgery (CS) has not been well characterized. This study used data from the National Inpatient Sample (NIS) to characterize AKI in this population and determine risk factors for postoperative AKI.
Materials and Methods: ACHD patients having undergone CS were identified in the NIS from 2005 to 2012. The presence of AKI was defined by ICD-9 codes during the included admission was then identified. Regression analysis was done to determine factors associated with AKI and determine if AKI impacts length of stay and cost of stay.
Results: 178,191 admissions were analyzed, 25,079 (14.1%) were documented as having AKI. AKI was associated with increased length of stay (21.6 vs 9.7 days, p < 0.001), cost of stay ($312,912 vs. 149,614, p < 0.001), and increased inpatient mortality (15.8% vs. 2.6%, p < 0.001). Surgeries associated with increased AKI frequency were heart transplant (OR 5.531, 95% CI 5.004 to 6.112), septal defect repair (2.103, 1.910 to 2.315), valvuloplasty with (2.017, 1.901 to 2.140) or without valve replacement (1.630, 1.544 to 1.720). Other factors associated with increased frequency of AKI by regression analysis included African American race, heart failure, and tachyarrhythmia.
Conclusion: AKI in those with ACHD occurs in at least 14.1% of admissions with CS based on coding in an administrative database. Specific interventions and risk factors are associated with increased risk of AKI. AKI increases length and cost of stay, and inpatient mortality. Understanding these findings may help improve surveillance and target interventions for AKI in those with ACHD undergoing CS.

O2215 - INCIDENCE AND 30 DAY MORTALITY OF PNEUMONIA RELATED HOSPITALIZATION IN CONGENITAL HEART DISEASE ADULTS COMPARED WITH THE GENERAL POPULATION A NATIONWIDE COHORT STUDY

Nicolas Madsen[1], Søren Paaske Johnsen[2], Henning Bækgaard Laursen[2], Morten Olsen[2]
Cincinnati Children's Hospital, Pediatrics, Cincinnati-United States[1]; Aarhus University, Clinical Epidemiology, Aarhus-Denmark[2]

Background/Hypothesis: The aging adult congenital heart disease (CHD) population may be vulnerable to pulmonary morbidity. We aim to study if adults with CHD are at increased risk for pneumonia (PNA) related hospitalization compared to the general population, and if this risk impacts the subsequent 30-day mortality.
Materials/Methods: This cohort study utilized nationwide population-based medical databases to identify individuals diagnosed with CHD in Denmark between 1963-1996. CHD subtype was divided by complexity: mild, moderate, severe, and univentricular. Adults, alive at age 15 years, were followed between 1977-2013 for the primary outcome (first hospital-based diagnosis of PNA) using the Danish National Registry of Patients (hospital discharge registry covering all Danish hospitals). For each CHD subject, we identified 10 controls from the general population, matched by sex and birth year. A unique personal identifier assigned at birth enabled virtually complete follow-up for migration, death or primary outcome. We computed cumulative incidences and hazard ratios (HR) of PNA-related hospitalization with stratification by CHD complexity. We compared the 30-day mortality associated with these PNA-related hospitalizations.
Results: We identified 13,997 CHD subjects. By 70 years of age, the cumulative incidence of first PNA-related hospitalization in CHD adults was 27%. The overall HR of PNA among CHD subjects compared with controls was 2.6 (95% CI: 2.4-2.8) [mild and moderate CHD: 2.3 (95% CI: 2.1-2.5), severe and univentricular CHD: 3.3 (95% CI: 2.9-3.8)]. Among CHD adults, the 30-day mortality for first PNA-related hospitalization was 9% (n = 1115). The corresponding birthyear and sex adjusted HR comparing 30-day mortality between CHD adults and controls was 1.2 (95% CI: 0.9-1.5) for subjects above 60 years of age, and 1.6 (95% CI: 1.1-2.3) for those under 60 years of age at time of PNA.
Conclusion: The incidence of PNA-related hospitalization, and 30-day mortality, were increased in CHD adults compared with the general population.

O2285 - SOLID NEOPLASMS IN FONTAN PATIENTS REPORT OF TEN CASES IN A SINGLE CENTER

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Background: Fontan has a unique circulation without subpulmonic ventricle. Chronic elevation of central venous pressure, low cardiac output and mild cyanosis is characteristic feature. Hepatocelluar carcinoma (HCC) and pheochromocytoma (PHEO)/paraganglioma (PG) has been reported recently in this population, but clinical data for solid tumor is scarce.
Method: We conducted a retrospective study on solid tumor developed in Fontan patients at Seoul National University Children's Hospital.
Results: Ten Fontan patients were diagnosed as solid tumor. Six patients (5 males and 1 female) were diagnosed as PHEO/PG and 5 patients (2 males and 3 females) as HCC. One patient had dual primary neoplasms, HCC and PHEO. Median age at diagnosis was 21.2 years (range, 13.7-35.4) in PC/PG and 27.0 years (range, 16.9-37.6) in HCC. Follow-up period after Fontan operation until development of tumor was 18.3 years (range 8.8-27.6) and 21 years (range 15.1-28.8), respectively. Clinical presentation to diagnosis of PHEO/PGL was hypertension in 2 patients and another 4 patients were incidentally detected by CT scan taken for other reasons. HCC was detected on routine screening abdominal sonography or CT in all patients. There were 2 mortalities in these patient group. One patient with malignant PHEO had multiple bone metastasis and died 5 years after the diagnosis due to heart failure and severe cyanosis. One patient with HCC had multiple lung metastasis at diagnosis and died from massive esophageal variceal bleeding in 2 months.
Conclusion: Solid tumor is another late complication after Fontan operation and it can become highly malignant. We have to be alarmed for the malignancy and check regularly for early detection and treatment of the tumor in Fontan patient group.

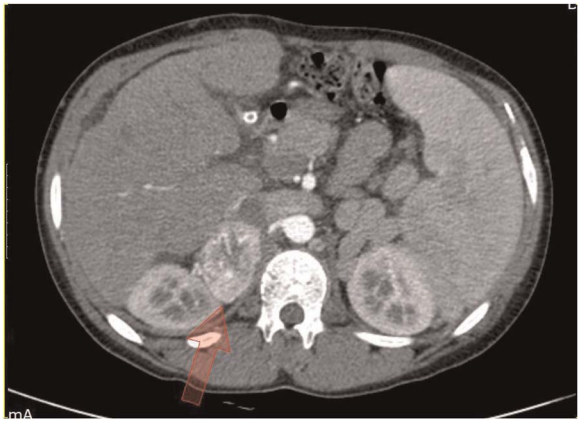

Figure 1.

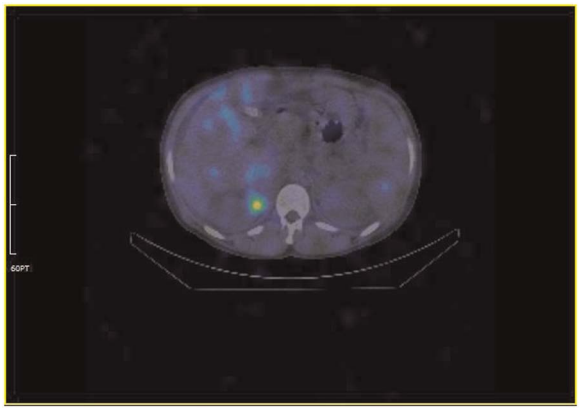

Figure 2.

O2459 - INDICATIONS AND MIDTERM OUTCOMES OF GERIATRIC CONGENITAL CARDIAC SURGERY A MULTICENTER STUDY

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Background: There is a paucity of data regarding surgical treatment of congenital heart defects in patients aged 60 years and older. The purpose of this study was to examine the surgical indications and midterm outcomes in this patient population.
Methods: Clinical and echocardiographic data were retrospectively gathered from a surgical database of adult congenital heart disease (ACHD) in two tertiary academic centers. Patients with isolated congenital aortic valve disease were excluded from this analysis. The median follow-up was 26 months (interquartile range 3-55 months).
Result: Between 2000 and 2015, 91 consecutive patients (mean 67 ± 5 years; range 60–82 years) underwent surgery for a congenital cardiac defect. Thirty-seven patients (41%) were in NYHA class $\geq$ III/IV. The mean preoperative EuroSCORE II was $4.1 \pm 6.1\%$. Nineteen patients (21%) had undergone previous cardiac surgery. Cardiac diagnoses included sinus venosus atrial septal defect (ASD) in 24 patients, secundum ASD in 23, atrioventricular septal defect in 13, pulmonary stenosis/Tetralogy of Fallot in 12, Ebstein anomaly in 7, coarctation in 2, coronary anomalies in 2, and miscellaneous defects in 8. Preoperative echocardiography revealed moderate tricuspid regurgitation (TR) in 30 patients (33%), and severe TR in 20 (22%), with a mean systolic pulmonary artery pressure (sPAP) of 44 ± 15 mmHg. Operative mortality occurred in 3 patients (4%). The median ICU stay was 2 days (interquartile range 1–4 days). Perioperative morbidity is depicted in Figure 1. At last follow-up, only 2 patients were in NYHA functional class $\geq$ III/IV. Follow-up echocardiography showed residual moderate TR in 11 patients, severe TR in 3, with a mean sPAP of 35 ± 13 mmHg.
Conclusions: The majority of our geriatric ACHD patients underwent surgery for either shunt lesions or right heart valve lesions. Our data demonstrate that surgery can be performed safely in this patient population. Surgical therapy led to improvement in functional status, sPAP and TR at intermediate follow-up.

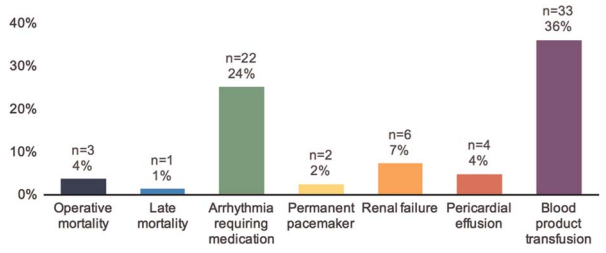

Figure.
Perioperative clinical outcomes.

O2475 - PATTERN OF CYANOTIC CONGENITAL HEART DISEASE IN ADULTS AT TERTIERY CARDIOVASCULAR CENTER IN INDONESIA

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Background: Grown-up patients with congenital heart disease (GUCH) often present difficult and challenging problems. In developing countries, a lot of children with congenital heart disease (CHD) reach adulthood without correction, due to delay in detection, limited resources, lack of knowledge and financial supports.
Objectives: This study was performed to evaluate the pattern of cyanotic CHD among Indonesian adults in Indonesia.
Methods: Retrospective study was conducted at National Cardiovascular Center Harapan Kita Hospital, Jakarta, as the tertiary cardiovascular center in Indonesia. Data was retrieved from the hospital database from 2007 to 2012.
Results: Among 817 GUCH, there were 104(12.7%) patients diagnosed as cyanotic CHD with median age of 20.9(17.0-66.3) years, 58(55.8%) female and 46(44.2%) male. Tetralogy of Fallot was found in 50(48.1%) patients, Ebstein's anomaly in 9(8.7%) patients, pulmonary atresia with ventricular septal defect in 9(8.7%) patients, complete atrio-ventricular septal defect in 7(6.7%) patients, double outlet right ventricle in 6(5.8%) patients, congenitally corrected transposition of the great arteries in 4(3.8%) patients, total anomalous pulmonary venous drainage in 4(3.8%) patients, transposition of the great arteries with ventricular septal defect in 2(1.9%) patients, and tricuspid atresia in 2(1.9%) patients. There were 5(4.8%) cases admitted with infective endocarditis, where 2 complicated with cerebral abcess and 1 with ischemic stroke. Arrythmia found in 4(3.8%), ventricular dysfunction in 7(6.7%), hematological disorders in 11(10,6%), renal dysfunction in 2(1.9%), and neurological complications in 3(2.9%) patients. Surgical management was performed in 52(50%) patients, whilst conservative treatment in 52(50%) patients. Blalock–Taussig shunt performed in 9(8.7%), tetralogy of Fallot repair in 32(30.8%), bidirectional cavopulmonary shunt in 4 (3.8%) and fenestrated Fontan in 4(3.8%) patients. Overall hospital mortalities were 3(2.9%) patients.
Conclusions: Cyanotic GUCH remains a complicated and unresolved health problem in Indonesia. Early detection and management during childhood is important to reduce the complications of this disease.

O2521 - CHALLENGES IN SURGICAL CORRECTION OF ADULT TETRALOGY OF FALLOT OUTCOMES WITH TARGETED CARE

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Background and Aims: Unrepaired adult Tetralogy of Fallot (TOF) is not uncommon in developing countries. Published literature reports discharge mortality of 5 - 15%. Our discharge mortality from 1995- 2005 was 2/81 (2.5%). Risk factors for death were - severe polycythemia, RV dysfunction and collaterals. Subsequently, following strategies were instituted : 1) Aggressive search and preoperative intervention for collaterals 2) staging BT shunt for persistent severe thrombocytopenia 3) Multiple preoperative phlebotomies 4) preoperative neuroimaging to detect infarcts and bleeds 5) Controlled RVOT resection 6) Preemptive management of RV dysfunction. Our aim is to assess whether these strategies resulted in improved outcomes.
Patients and Methods: From 2006 to 2016, total 103 patients (Median age 21 years, range 16 - 50) with diagnosis of TOF (n = 94), DORV [TOF type] (n = 5), TOF with Absent Pulmonary valve (n = 4) were operated . Preoperative MAPCA coiling was required in 21 and 28 received multiple preoperative phlebotomies. 52 patients received transannular patch,5 required pulmonary valve replacement, 2 tricuspid valve repair, 1 aortic and 1 mitral valve replacement . 4 underwent a staging BT shunt for persistent severe thrombocytopenia. Median preoperative hematocrit value was 55% (range 34-78).
Results: In hospital mortality was 0/103 with no late deaths. Median duration of ventilation, ICU and hospital stay was 17 hours (range 4-192), 42 hours (range 8-258) and 8 days(range 4-25) respectively. 5/103 underwent reexploration for bleeding and one required multiple craniotomies for recurrent subdural hematoma. 2 underwent subsequent pulmonary valve replacement after 5 and 8 years.
Conclusions: Early outcomes after surgery for adult TOF can be greatly improved with highly individualized management despite multiple perioperative challenges.

O2525 - ARTERIAL SWITCH OPERATION FOR PATIENTS WITH TRANSPOSITION OF THE GREAT ARTERIES A SINGLE CENTRE TWENTY EIGHT YEAR EXPERIENCE

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Background: Complete transposition of the great arteries (D-TGA) is a life-threatening malformation in neonates which requires immediate management. We sought to evaluate our long-term results with arterial switch operation (ASO) for D-TGA during the last 30 years.
Methods: We collected data on 260 patients with D-TGA who underwent arterial switch operation (ASO) between January 1987 and December 2016. Outcome included survival, need for late re-operations/re-intervention and clinical status.
Results: Original diagnoses included: simple D-TGA (n = 177, 68%) and complex D-TGA (n = 83, 32%). The percentage of cases of D-TGA diagnosed prenatally has increased during the last 10 years ($p < 0.0001$). Median age at ASO was 8 days (IQR 6-12 days). Fifteen patients (5.8%) died in hospital (6/177 simple D-TGA, 3.3% and 9/83, complex D-TGA, 10.8%). Sixty-eight patients (26%) presented post-operative complications including: low output syndrome (n = 20, 7.7%), respiratory complications (n = 20, 7.7%) and arrhythmias (n = 18, 6.9%). Median follow-up time after ASO was 10 years (IQR 3.7-17.2 years). There were 2 (2/245 survivors, 0.8%) late deaths. Qverall survival probability at 10 and 20 years after ASO was 93.4% and 92.4%, respectevely. Thirty-five patients (14%) required re-operations/re-interventions, mainly for right ventricular outflow tract obstruction (n = 28). Freedom from re-operation/re-intervention at 10 and 20 years after ASO was 87.2% and 77.8%, respectevely. Prenatal detection of D-TGA reduced the probability of early and late mortality ($p = 0.04$ and $p = 0.04$) and of post-operative complications ($p = 0.04$).
Conclusions: The ASO for D-TGA can performed with low early and late mortality. The prenatal detection of D-TGA reduces significantly postoperative mortality and morbidity.

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O2766 - QUALITY OF LIFE AND EXERCISE CAPACITY IN CONGENITAL HEART DISEASE PATIENTS WITH PROSTHETIC HEART VALVES

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Introduction: Most studies concerning prosthetic valve replacement in congenital heart disease have a focus on the functionality of the prosthesis and mortality. The increased life expectancy of this congenital population shifts the focus to their quality of life, which becomes more and more important. The aim of this study was to find predictors of quality of life and exercise capacity.
Materials and Methods: In a multi-center cross sectional observational study of adults with congenital valve disease, patients with a pulmonary valve replacement (n = 142) were compared with those with an aortic valve replacement (n = 153) and patients with both aortic and pulmonary valve replacement (n = 54). To compare quality of life the Dutch version of the Rand-36 questionnaire was used. Exercise capacity was tested using a treadmill or bicycle ergometer.
Results: Patients receiving a pulmonary valve had significantly lower bodily pain scores than those receiving an aortic valve 82.0 ± 23.2 vs 89.9 ± 16.3 (p = 0.03). On all other categories, patients receiving a pulmonary valve also scored higher, although not significantly when corrected for age and gender. Exercise capacity was a significant predictor for physical functioning ($p < 0.001$), physical role functioning ($p = 0.018$), bodily pain ($p = 0.001$), general health ($p = 0.004$) and social functioning ($p = 0.04$), of the quality of life questionnaire. Patients with pulmonary valve prosthesis had significant lower exercise capacity than those receiving an aortic prosthesis 77.4 ± 2.1 vs 87.7 ± 2.3 ($p < 0.001$). Being employed and playing sports are both predictors for exercise capacity (p-0.002, p = < 0.001).
Conclusion: Even though the patients receiving a pulmonary valve replacement had significantly lower scores on exercise capacity testing compared to patients with aortic and double valve replacement, paradoxically they scored equally or better on the quality of life questionnaires.

O2845 - PERI OPERATIVE MANAGEMENT OF HEART TRANSPLANTATION IN ADULT FAILING FONTAN PATIENTS

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Background: Heart transplantation for late failure of the Fontan circulation is high risk. Anaesthetic challenges relate to anatomic complexity, previous cardiac surgery and unique physiology.
Objective: We describe the perioperative management of adult failing Fontan patients undergoing heart transplantation and identify strategies that may improve outcome.
Methods: A retrospective case-note analysis of adult Fontan heart transplants at Freeman Hospital, Newcastle upon Tyne, UK between 2009 and 2016 was performed.
Results: 22 patients (26 (17-43) years, 60.1 ± 9.4 kg) underwent isolated heart transplant for failing Fontan. 16 remained in hospital for optimisation including milrinone infusion in 14. Surgical strategy involved pulmonary artery and bicaval anastamosis reconstruction. Knife-to-skin to cardiopulmonary bypass (CPB) was 116.7 ± 51.2 minutes. CPB time was 275 ± 169.1 minutes and organ ischaemic time 156 ± 58.5 minutes. CPB strategy included supranormal flows, maintained haematocrit of 0.30, mannitol and colistin in the prime (n = 14) and fresh frozen plasma (n = 18). Separation from CPB was with epinephrine, milrinone, vasopressin and isoprenaline with 15 requiring inhaled nitric oxide and 18 pacing. All received platelets, 21 had continuous aprotinin and seven cryoprecipitate. Extubation was at 4 (0-56) days. Three required tracheostomy. ICU stay was 11 (2-77) days. 18 required renal replacement therapy for 16.5(4-132) days. One required late renal transplant. Mortality was 31.8%: peri-operative bleeding and multi-organ failure (4), bowel and lower limb ischaemia (2) and sudden cardiac arrest at 132 days (1). ECMO was used post-operatively in five: failure to wean from CPB (1), early graft failure (3), sudden late collapse (1). Two survived with major complications (bilateral above knee amputation, kidney failure, intracranial haemorrhage). There was no allograft rejection.
Conclusion: Heart transplantation for failing Fontan remains a high-risk procedure. Successful management requires meticulous surgical technique, novel bypass management and strategy for coagulopathy. A multidisciplinary team approach is key.

O2924 - SYSTOLIC BLOOD PRESSURE RESPONSE AND ARRHYTHMIA DURING EXERCISE PREDICT PREGNANCY OUTCOME IN WOMEN WITH CONGENITAL HEART DISEASE.

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Background: Low New York Heart Association functional class (NYHA) is associated with poor outcome of pregnancy in women with congenital heart disease (WCHD). However, there have been no established criteria for the safer pregnancy in WCHD.
Purpose: To identify major risk factors and determine referral cardiovascular variables during cardiopulmonary exercise testing (CPX) in WCHD.
Method and Results: We retrospectively reviewed 68 WCHDs who had undergone cardiopulmonary exercise testing (CPX) within 5 years before their delivery and the pregnancy outcomes were compared with NYHA, CPX-derived variables (heart rate, oxygen uptake, systolic blood pressure (mmHg) at peak exercise [P-HR, P-VO2, and P-SBP, respectively]) and clinically relevant arrhythmia during CPX (Ex-Arr). Seventeen maternal cardiac and 26 neonatal events occurred. All variables, were associated with maternal cardiac and neonatal events ($p < 0.05$-0.001). Of these, P-SBP and Ex-Arr were the independent determinants of the maternal (odds ratio [OR]:0.96, 95% confidence interval [CI]: 0.92-0.99, $p < 0.05$ for P-SBP) and neonatal events (OR: 0.95,

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95% CI: 0.91-0.99, p < 0.01 for P-SBP, and OR: 21.2, 95% CI: 2.1-559, p < 0.01 for Ex-Arr), except for Ex-Arr for maternal events. The cutoff value of SBP for maternal and neonatal events was 150 and 154, respectively.
Conclusion: All major CPX-derived variables, especially, P-SBP and Ex-Arr, can predict adverse outcome during pregnancy in WCHD. P-SBP ≥ 150 (mmHg) without Ex-Arr could be a reliable reference value for safer pregnancy outcome in WCHD.

AMBULATORY

O1101 - COMPARISON OF INITIAL THERAPY OF INTRAVENOUS IMMUNOGLOBULIN VERSUS INTRAVENOUS IMMUNOGLOBULIN PLUS INFLIXIMAB IN KAWASAKI DISEASE PATIENTS PRESENTING WITH CORONARY ARTERY LESIONS

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Background: 80% of Kawasaki Disease (KD) children who develop coronary artery lesions (CAL) have them present at time of diagnosis. These children represent a high risk group that may benefit from more aggressive initial therapy. Infliximab has been shown to decrease inflammation in KD patients when added to standard therapy. Therefore, we compared intravenous immunoglobulin (IVIG) alone with IVIG plus infliximab as an initial treatment in KD patients with CAL to reduce IVIG resistance and to evaluate coronary artery changes between groups.
Methods: KD patients were retrospectively reviewed from 2009-2016. CALs were defined as left anterior descending artery and right coronary artery z-scores > 2.5. KD patients with CALs on initial echocardiogram who were treated with IVIG were compared with those treated with IVIG + infliximab. Clinical characteristics, length of stay, and additional therapy were compared using chi-squared and Fischer's exact tests. Effect of treatment on CALs between groups was assessed using linear mixed models at three time points: baseline, 2 weeks, and 6 weeks post treatment.
Results: 34 KD patients with CALs were treated with IVIG and 35 KD patients with CALs were treated with IVIG + infliximab. There were no differences in clinical and laboratory characteristics between the two groups (Table 1). 50% (17/34) of KD patients treated with IVIG required additional second therapy compared to 17% (6/35) of KD patients treated with IVIG + infliximab (p < 0.008). There was a non-statistically significant trend towards shorter length of stay in the IVIG + infliximab group. There were no significant differences between treatment groups for resolution of CALs or fall in CRP.
Conclusions: IVIG + infliximab should be considered as initial therapy in KD patients who present with CALs as both reduce the need for additional therapy.

Table 1. Patient Clinical and Laboratory Characteristics.

	IVIG (n = 34)	IVIG + Infliximab (n = 35)	p-value
Gender (male/female)	28/6	26/9	0.60
Ag ≥ 1 yearold	28 (82%)	22 (63%)	0.06
Age < 1 year old	6 (18%)	13 (37%)	0.11
Complete Symptoms (≥4)	21 (62%)	19 (55%)	0.59
Day of illness treated	8.09 ± 4.26	9.29 ± 6.69	0.37
Admit to PICU	(15%)	6 (17%)	1.00
CRP	12.52 ± 8.81	13.43 ± 11.23	0.71
ESR	64.94 ± 29.52	58.71 ± 28.05	0.38
WBC	15.24 ± 5.45	15.21 ± 4.54	0.98
Hgb	11.60 ± 1.57	10.95 ± 1.79	0.12
Platelets	364.71 ± 113.62	429.43 ± 170.13	0.07
Albumin	3.31 ± 0.65	3.32 ± 0.63	0.92
AST	55.03 ± 50.25	51.99 ± 39.91	0.79
ALT	57.03 ± 56.89	53.79 ± 35.03	0.78
GGT	78.12 ± 66.00	116.31 ± 89.13	0.33
Urine WBC ≥10	3 (9.68%)	4 (12.90%)	1.00
Length of stay Mean ± SD	5.94 ± 6.33	3.86 ± 2.16	0.07

O1228 - INFLUENCE OF CARDIAC INTERVENTION ON NEURODEVELOPMENT IN YOUNG CHILDREN WITH CONGENITAL HEART DISEASE IN CENTRAL SOUTH AFRICA THREE MONTH AND SIX MONTH OUTCOMES

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Background: Congenital heart disease (CHD) survivors are at risk of neurodevelopmental morbidity. The neurodevelopmental outcomes of children with CHD in South Africa are largely unknown. The aim was to determine the neurodevelopmental outcome of young children with CHD following a cardiac intervention.
Materials and Methods: Forty-eight children, 30 months and younger, were recruited into this observational descriptive study. Children who had previous or emergency cardiac surgery were excluded. Development was assessed using the Bayley Scales of Infant and Toddler Development, Third Edition (BSID-III) before, at three-month and six-month post-cardiac intervention. Developmental outcomes were compared over time, and variables associated with developmental performance determined.
Results: Baseline data was collected for 40 children. The majority of children (n = 26) underwent open-heart surgery in infancy with cardiopulmonary bypass. Most children (n = 30) had moderate disease severity, with 20% (n = 8) having cyanotic lesions. A quarter of the children (n = 10) had Down syndrome (DS). Motor delays were prevalent (27.5%) prior cardiac intervention. Motor performance improved, but language and cognitive performance declined post-cardiac intervention, with age and increasing skill complexity. Hypotonia presented in 45% of children prior to cardiac intervention, resolving post-cardiac intervention in all children without DS. Fifty-nine percent of the children were at-risk of, or presented with developmental delays; 42.5% requiring occupational therapy, 55% physiotherapy and 59% speech therapy services. There was no significant change in the developmental outcome over time. BSID-III subscale scores remained below the test mean of 100. The presence of DS (p < 0.001), disease severity (p = 0.02), maternal age (p < 0.001), age at first surgery (p <0.01), and growth prior to cardiac intervention (p = 0.04) were significantly associated with developmental performance. Social disadvantage negatively impacted developmental performance.

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Conclusions: Neurodevelopmental delays prior to, and post cardiac intervention were prevalent in this study sample. The majority of the children were at risk, or had developmental delays requiring early referral to therapeutic services.

O1319 - IMPROVING PATIENT ACCESS AND SATISFACTION WITH INDEPENDENT PEDIATRIC NURSE PRACTITIONER CARDIOLOGY CLINICS

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Background: In 2008, wait times for 3rd to next available appointment exceeded 40 days in the cardiovascular outpatient department at a children's hospital. To address the gap in available appointments, Independent Pediatric Nurse Practitioner Cardiology Clinics were implemented as a strategic solution to improve patient access to urgent and non urgent appointments.
Methods: A sample of 128 patients who presented concurrently in physician or pediatric nurse practitioner cardiology clinics from December 2009 through February 2010 was recruited for participation. The hospital's ambulatory patient satisfaction survey was utilized to measure level of patient satisfaction. Survey responses were evaluated using Fisher's exact test. Appointment wait times were compared pre and post implementation of pediatric nurse practitioner clinics.
Results: There was no statistically significant difference in patient satisfaction outcomes between physician or nurse practitioner clinic type. Appointment wait time decreased from 46 to 4 days. Pediatric nurse practitioner clinics included a statistically higher percentage of urgent appointments compared to physician clinics.
Conclusions: This innovative health care delivery model helped reduce appointment wait times while earning high levels of patient satisfaction. Independent Pediatric Nurse Practitioner clinics exemplify the Institute of Medicine's goals for advanced practice nursing and epitomizes what contemporary nursing practice can achieve in a Quaternary care and academic center of excellence. Cardiovascular Nurse Practitioners support the mission and vision of Heart Center to provide timely, patient-centered, cost effective, expert cardiovascular care.

O1638 - EXTENDING PAEDIATRIC CARDIOVASCULAR CARE TO GULU REGIONAL REFERRAL HOSPITAL IN UGANDA CHALLENGES AND OPPORTUNITIES IN A FOUR YEAR EXPERIENCE

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Background: Pediatric cardiac services in resource limited countries are often found only in big cities. Because of these inequalities in access, many more rural children with heart disease suffer death or disability compared to those living near cities. We describe the start a satellite cardiac clinic at Gulu Regional referral Hospital (GRRH) in Gulu, a northern Uganda City 335 km from the capital Kampala.
Methods: A pediatric cardiologist was trained at the Uganda Heart Institute (UHI) in Kampala from September 2010 to August 2013. Initial challenges included lack of functional diagnostic equipment. A RHD primary school screening program in Gulu began in October 2013 through a Collaboration between the UHI and Children's National Health System, USA which provided a portable echo machine. A satellite cardiac clinic, staffed by a Paediatric cardiologist, two research nurses, and administrative support was established at GRRH with support from RHD Action Uganda to provide care for RHD and other cardiac patients (children and adults) from the entire northern region.
Results: Total outpatient visits increased from 20 in 2013 to over 1400 in 2016. Total echoes performed over the period for children age <18 years were 728 (Normal = 263; CHD = 217; Acquired heart disease = 248, majority being 214 RHD drawn from both school screening and symptomatic cases). Twenty children have been linked to surgical or catheter-based care at UHI (n = 15) or abroad (n = 5). Monthly outreach visits are also conducted at Lira Regional Referral Hospital. Local and international partnerships have evolved for care delivery and research in Gulu. Linkage of more patients for definitive care is most challenging for patients who cannot afford transport to UHI.
Conclusions: Extending pediatric cardiac care to regional hospitals in Uganda leads to improved care delivery. However, more resources are needed to improve access to tertiary care that is only available in Kampala.

O1679 - PROMOTING PHYSICAL ACTIVITY TO CHILDREN WITH CONGENITAL HEART DEFECTS IDENTIFYING IMPORTANT KNOWLEDGE TRANSLATION GAPS

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Background/Hypothesis: Physical activity (PA) counselling during clinical encounters is recommended because children with congenital heart defects (CHD) lead sedentary lives, decreasing quality of life and increasing morbidity risk. Clinicians encourage active lifestyles but families remain uncertain about PA. This study sought to identify PA knowledge translation needs, hypothesizing they would differ between CHD children, parents, and recreation/healthcare clinicians.
Materials and Methods: Interviews/focus groups with 13 CHD patients (6 female), 22 parents (18 female), 14 recreation professionals (9 female), and 24 paediatric cardiology healthcare specialists (18 female). Audio-recorded discussions were transcribed verbatim for inductive qualitative analysis within pre-defined coding frames: a) activity barriers/facilitators, and b) practice recommendations.
Results: All study participants identified a need for written PA resources from a recognized source. "I wish there was something that answered all of the basic, common questions", using specific, easy-to-understand language. "The word cardiac scares people" was also a shared perspective, but the attributed sources of fear and perceived impacts differed. Parents reported instructors being fearful of their children. Recreation leaders felt parents and doctors feared their cardiac PA expertise was limited. Healthcare specialists reported parent fears override their PA recommendations.

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Perceptions of successful PA differed between study groups. Children, parents and recreation leaders emphasized fun, engaging activities with family/friends, practical advice on how to make PA happen and novel activity ideas that suit cardiac limitations. Encouraging lots of PA experience/practice and consistently providing/repeating information was recommended. Healthcare providers felt PA adaptations for each child's abilities "may be a dream", encouraging alternate roles (e.g., referee) if needed.
Conclusions: All participants agreed cardiac fear is a major PA barrier, but differed regarding fear sources and impacts. Revising healthcare practice to focus on encouraging PA participation, via practical suggestions and clear information from credible sources, and enhancing awareness of recreation options and expertise are recommended.

O1788 - ANALYSING CURRENT PRACTICE IN THE ASSESSMENT OF PAEDIATRIC CHEST PAIN

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Background: Chest pain is a common presenting complaint to General Practitioners, Paediatricians and the Emergency Department, often resulting in paediatric cardiology referral. However, unlike adult chest pain, paediatric chest pain rarely has a cardiac aetiology, often resulting in high levels of unnecessary testing and associated anxiety. Currently there is no UK guidance for paediatric chest pain assessment, which may be a contributory factor for variation in clinical practice.
Aims: To assess the outcome of referrals to a paediatric cardiology department with primary symptom of chest pain.
Methods: A cross-sectional observational study of paediatric patients aged 4-18 years referred to the paediatric cardiology department of a regional tertiary referral centre, with the primary symptom of chest pain. Prospective and retrospective data was gathered in paediatric cardiology clinics and from patient notes between October-December 2016. Data was collected regarding presenting symptoms, history, examination, diagnostic testing and eventual diagnoses.
Results: 42 patients were included in the study (26 retrospective, 16 prospective). 25 patients were female and 17 male, with an average age of 11.47 years (+/- 3.48). Chest pain aetiologies included: 55% (23) idiopathic, 31% (13) musculoskeletal, 7% (3) gastrointestinal, 2% (1) anxiety. 2 patients were still undergoing testing, but their chest pain was not attributed to a cardiac pathology. No patients were found to have cardiac chest pain. All patients had an electrocardiogram (ECG) and echocardiogram and each had an average of 2.9 diagnostic tests. Additional tests included exercise stress tests and ambulatory ECG monitoring.
Conclusions: Paediatric chest pain rarely has a cardiac aetiology. Practice variation and unnecessary resource use are a concern, and a standardised approach to assessment could improve patient care by limiting unfruitful cardiac testing and associated anxiety. A detailed history, physical examination and ECG are usually sufficient for diagnosis of the cause of paediatric chest pain.

O1881 - MATERNAL OPIOID AGONIST THERAPY AND RISK OF CONGENITAL HEART DISEASE IN THE FETUS

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Background: Maternal opioid use disorder (OUD) during pregnancy has increased over the past decade; current standard of care is treatment with methadone (MTD) or buprenorphine (BPH). Frequently there is concurrent use of psychiatric medications, illicit drugs or cigarettes. Maternal analgesic opioid use early in gestation has been associated with congenital heart disease (CHD). Little data exists about the association of opioid agonist therapy (OAT) with MTD or BPH during pregnancy and risk for CHD.
Objective: Assess impact of opioid agonists for OUD during pregnancy on incidence of CHD.
Methods: Retrospective cohort study of 744 mother-infant dyads at Boston Medical Center between 2006-2015. Inclusion criteria were infants >36 weeks gestational age (GA) and mothers on MTD or BPH at delivery. Outcomes included diagnosis of CHD as per the electronic medical record. Maternal and infant demographics, and pregnancy co-exposures were analyzed. Analysis performed included chi- square test for categorical and dichotomous variables and simple logistic regression for continuous variables.
Results: Of 744 infants, 454 (61.0%) were MTD and 290 (39.0%) BPH exposed. OAT was associated with an increased incidence of CHD (Table) of 2.54% as compared to 0.8% in the general population. There was no difference in CHD based on MTD vs BPH exposure. Earlier first prenatal visit and GA at delivery, or older maternal age trended toward increased risk of CHD, but did not meet significance. Use of psychiatric medications and smoking did not increase risk of CHD. Illicit drug use during the third trimester (n = 61, 8.2%) was associated with increased CHD ($p = 0.01$). In a subgroup analysis excluding those with illicit drug exposure, the incidence of CHD remained higher than the general population at 2.01%.
Conclusion: Prenatal use of opioid agonists for treatment of OUD is associated with increased risk of CHD, with no differences in risk between MTD and BPH.

Table.

Congenital heart defect	N
Ventricular septal defect	12
Atrial septal defect	1
Pulmonary Valve Stenosis	1
Ebstein Anomaly	1
Cardiomyopathy	1
Endocardial Cushion Defect	1
Venous Malformation	2
Bicommissural aortic valve	1

O1931 - EFFECTS OF US STATE NEWBORN CRITICAL CONGENITAL HEART DISEASE SCREENING POLICIES ON INFANT CARDIAC DEATHS

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Background: With the goal of reducing preventable deaths among infants with critical congenital heart disease (CCHD), screening for CCHD using pulse oximetry has been recommended in the

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United States since 2011. Incorporating this recommendation as part of mandated newborn screening policy, however, is up to each individual state. This study aimed to estimate the effects of state CCHD newborn screening policies on infant cardiac deaths from congenital heart disease (CHD) in the United States.
Methods: Using 2007-2013 period linked birth and infant death data, we utilized a difference-in-differences strategy to estimate the impact of state CCHD screening policies on early infant (24 hour– < 6 months of age) mortality rates for CCHD or other/unspecified CHD. We adjusted for state trends and state-period-specific variables and projected avoidable deaths at the national level.
Results: Between August 31, 2011 and June 1, 2013, nine states implemented mandatory CCHD screening policies. In these states, adjusted rates of early infant mortality from CCHD decreased by 36.8% (95% CI: 12.5%–54.4%) for births during months after implementation of mandatory CCHD screening compared to prior periods and states without CCHD screening policies. Other/unspecified CHD adjusted mortality rates decreased by 17.6% (95% CI: 1.6%–30.9%). We projected that universal CCHD screening in the United States could reduce deaths due to recognized CCHD by 42-180 per year, relative to not screening for CCHD, not including potential reduction in infant cardiac deaths with non-critical or unspecified CHD codes, some of which may reflect undiagnosed CCHD.
Conclusions: These results suggest that mandatory CCHD screening policy achieves the goal of reducing preventable deaths and is an effective public health intervention for the prevention of infant cardiac deaths. These results have important implications for jurisdictions considering CCHD screening policies.

O2038 - EFFECTIVENESS AND SAFETY OF STATIN THERAPY IN CHILDEN A REAL WORLD CLINICAL PRACTICE EXPERIENCE

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Background: Existing evidence of statin use in children is predominantly limited to short-term research trials. To evaluate effectiveness and safety of longer-term statin use in clinical practice, we reviewed results of all children treated for >6 months (m) from 5 Pediatric Lipid Clinics.
Results: There were 334 patients who had 2197 clinical assessments. In 84%of patients, the diagnosis was familial hypercholesterolemia; 16% had combined hyperlipidemia. Mean age at statin initiation was 12.9 ± 3.0 years. Mean duration of therapy was 3.2 years (IQR: 1.6-4.6) with 46% over 3 years. Initial prescribed statin varied over time but 65% were started on atorvastatin and 78% remained on the first statin prescribed. Mean compliance was clinically assessed at 92% and 330 patients(99%) remained on statin therapy at review end. The table displays results for regression analyses adjusted for repeated measures over time for two intervals: baseline(B/L) to 6 m post statin initiation, and 6 +/-3m post initiation to last follow-up(F/U). Statin treatment was initially associated with significant reductions in TC, LDL-C and non-HDL-C with no significant change in fasting TG, HDL-C or safety labs. There were further significant reductions in TC and LDL-C with longer treatment. Only 6 patients complained of any symptoms, none attributed to treatment. While 11% of patients had transient AST or ALT > 3xULN or CK > 10xULN during follow-up, none required discontinuation of statins for persistent elevation. Mean fasting glucose was unchanged throughout F/U and no patient developed diabetes mellitus. Longer duration of statin therapy was not associated with increased odds of lab abnormalities.
Conclusion: These findings in a large series of pediatric patients from real-world clinical practice show that statin therapy for hypercholesterolemia is safe and effectively lowers LDL-C on longer-term follow-up. Side effects are rare and discontinuation of treatment is uncommon.

	BASELINE (B/L) Mean (IQR)	6 +/ -3 mos Mean (IQR)	LAST F/U Mean (IQR)	Change/yr, B/L to 6 mos Mean (95%CI)	Change/yr, 6 mos to last F/U Mean (95%CI)
Total Cholesterol (mg/dL/ mmol/L)	267 (240-317) 6.9 (6.2-8.2)	228 (190-265) 5.9 (4.9-6.9)	207 (178-239) 5.4 (4.6-6.2)	-34.8 (-55.4; -14.2) (p < 0.001)	-11.1 (-12.9; -9.3) (p < 0.001)
LDL-C (mg/dL/ mmol/L)	200 (167-246) 5.2 (4.3-6.4)	157 (116-195) 4.1 (3.0-5.0)	140 (110-168) 3.6 (2.8-4.3)	-35.0 (-54.1; -15.9) (p < 0.001)	-10.7 (-12.3; -9.1) (p < 0.001)
HDL-C (mg/dL/ mmol/L)	45 (39-53) 1.2 (1.0-1.4)	46 (39-55) 1.2 (1.0-1.4)	45 (38-54) 1.2 (1.0-1.4)	-2.0 (-6.7; 2.8) (p = 0.42)	-0.074 (-0.50; 0.35) (p = 0.73)
Triglycerides (TG) (mg/dL/ mmol/L)	100 (69-141) 1.1 (0.8-1.6)	93 (66-129) 1.1 (0.8-1.5)	91 (66-130) 1.0 (0.8-1.5)	-10.7 (-36.0; -14.4) (p = 0.40)	-1.38 (-2.85; 0.093) (p = 0.07)
Non-HDL-C (mg/dL/ mmol/L)	220 (193-268) 5.7 (4.9-6.9)	178 (141-220) 4.6 (3.7-5.7)	161 (130-193) 4.1 (3.4-4.9)	-34.8 (-62.6; -6.9) (p = 0.01)	-6.2 (-12.6; 0.30) (p = 0.06)
ALT (U/L)	24 (17-33)	24 (18-32)	25 (17-35)	3.1 (-2.7; 8.9) (p = 0.29)	0.55 (-0.092; 1.19) (p = 0.09)
AST (U/L)	26 (21-32)	27 (21-34)	24 (19-31)	2.3 (-2.6; 7.2) (p = 0.36)	0.69 (-1.25; -0.12) (p = 0.02)
CK (U/L)	95 (74-138)	99 (69-140)	94 (67-141)	75.5 (-45.7; 196.6) (p = 0.22)	9.9 (-5.0;24.7) (p = 0.19)
Fasting glucose (mg/dL)	86 (83-92)	86 (83-92)	88 (83-94)	11.6 (-15.9; 39.0) (p = 0.41)	0.60 (-0.17; 1.36) (p = 0.13)

O2084 - RIGHT VENTRICULAR DYSFUNCTION AND EXERCISE INTOLERANCE IN PATIENTS WITH MODERATE OR SEVERE PULMONARY REGURGITATION IN PATIENTS WITHOUT TETRALOGY OF FALLOT. ARE THEY THE SAME AS TETRALOGY OF FALLOT

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Background: Right ventricular (RV) remodeling and dysfunction secondary to pulmonary valve regurgitation (PR) are common after intervention(s) for congenital pulmonary stenosis (PS) or atresia and intact ventricular septum (PA/IVS). Pulmonary valve replacement has largely been evaluated in patients after repair of tetralogy of Fallot (TOF), but it is unknown whether PR has similar effects in on RV function in non-TOF patients. The objective of this study was to compare exercise tolerance and RV function in non-TOF versus TOF children with similar PR and RV dilation.
Methods: Thirty PS or PA/IVS children after intervention(s) with moderate or severe PR (non-TOF group) and 30 patients after surgical TOF repair (TOF group), matched for age and RV end-diastolic volume index (RVEDVi), measured by cardiac magnetic

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resonance (CMR) were retrospectively studied. Patients with residual PS (>25 mmHg) were excluded. Clinical data, ECG, exercise capacity, echocardiography and CMR were compared between groups.

Results: The groups were well matched for age, RVEDVi, gender distribution and CMR PR fraction (Table). All TOF patients had surgical repair of the RV outflow tract and had a right bundle branch block, versus 13% and 57% of non-TOF, respectively. QRS was wider in TOF patients. Parameters of RV function and exercise capacity were similarly reduced in both groups. The mean RV ejection fraction (RV-EF) and peak oxygen consumption (% predicted) in non-TOF versus TOF were: 48.7 ± 6.4% versus 48.5 ± 7.2%, p = 0.93; and 82.5 ± 17.7% versus 75.6 ± 20.4%, p = 0.24, respectively. Negative correlation was found in both groups between RVEDVi and RV-EF (non-TOF: r = -0.39, p = 0.04, TOF: r = -0.40, p = 0.03).

Conclusions: RV function and exercise capacity are similarly reduced in non-TOF and TOF patients with clinically significant PR matched by RV volume. These data suggest that RV dilatation, rather than the intrinsic disease, drives RV dysfunction. This implies that criteria for pulmonary valve replacement should be applicable to both groups.

Table. Demographic, clinical, ECG, exercise test, echocardiogram and CMR data in non-TOF versus TOF patients.

		Non-TOF	TOF	
	Patients analyzed	Mean ± SD or n (%)	Mean ± SD or n (%)	P Value (McNemar's or paired t-test)
Demographics and clinical data				
Gender: Male	30	19 (63.3%)	14 (46.7%)	0.36
Age (at CMR), years	30	13.9 ± 2.9	13.8 ± 2.9	0.93
Height, cm	30	158.7 ± 15.5	149.3 ± 21.1	0.02
Weight, kg	30	55.1 ± 19.3	53.7 ± 32.7	0.85
Body surface area, m^2	30	1.55 ± 0.34	1.45 ± 0.41	0.33
Pulmonary atresia	30	8 (26.7%)	3 (10%)	0.18
Surgical repair of the RVOT	30	4 (13.3%)	30 (100%)	<0.001
ECG (closer to CMR)				
QRS Width, ms	30	102.4 ± 20.9	136.8 ± 21.3	<0.001
RBB block presence	28	17 (56.7%)	28 (100%)	0.001
Exercise test, GXT (closer to CMR)				
PeakVO_2, ml/kg/min	21	36.8 ± 7.4	33.1 ± 9.4	0.12
Peak VO_2, predicted %	21	82.5 ± 17.7	75.6 ± 20.4	0.24
Echocardiogram (closer to CMR)				
Age (at echocardiogram), years	30	13.9 ± 3.4	13.8 ± 2.9	0.93
PR qualitatively				
Moderate	30	8 (26.7%)	2 (6.6%)	0.08
Severe	30	22 (73.3%)	28 (93.3%)	0.08
RVOT gradient, mmHg	30	11.8 ± 4.8	15.1 ± 5.5	0.03
RV-RA gradient through TR jet, mmHg	21	24.2 ± 5.3	29.7 ± 8.3	0.04
RV end diastolic diameter, cm	25	3.25 ± 0.55	3.34 ± 0.58	0.58
RV end diastolic diameter Z-score	24	3.7 ± 1.0	4.0 ± 1.0	0.39
RV end diastolic area index, cm^2	27	18.7 ± 4.3	22.4 ± 4.1	0.004
RV end systolic area index, cm^2	27	10.9 ± 3.2	12.9 ± 3.4	0.03
RV-FAC, %	28	41.4 ± 7.6	43.2 ± 5.6	0.3
Patients with RV-FAC <40%	29	11 (37.9%)	8 (27.6%)	0.55
LV ejection fraction, %	28	65.4 ± 7.5	65.2 ± 5.8	0.89
Cardiac magnetic resonance data, CMR				
PR fraction, %	30	32.2 ± 13.2	38.1 ± 12.1	0.11
RV end diastolic volume index, ml/m^2	30	162.2 ± 32.4	163.2 ± 33.3	0.91
RV end systolic volume index, ml/m^2	28	84.2 ± 24.2	86.4 ± 22.4	0.75
RV ejection fraction, %	30	48.7 ± 6.4	48.5 ± 7.2	0.93
Patients with RV- EF < 45%	29	8 (27.6%)	7(24.1%)	1.0
LV ejection fraction, %	30	56.6 ± 6.2	55.6 ± 7.1	0.62

Abbreviations (not mentioned in the abstract): LV: Left ventricle; RA: Right atrium; RVOT: Right ventricular outflow tract; SD: standard deviation; TAPSE: Tricuspid annular plane systolic excursion; TR: Tricuspid regurgitation; VO_2: Oxygen consumption.

O2102 - STATE OF THE ART CLINICAL CARE OF THE FONTAN CIRCULATION A MULTI INSTITUTIONAL SURVEY OF PRACTICE VARIABILITY IN IDENTIFIED FONTAN PROGRAMS ACROSS THE UNITED STATES

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Background: Significantly more children with single ventricle physiology are surviving past Fontan surgery. This unnatural circulation imposes pervasive effects on growth and development and has unique modes of failure. Programs specifically designed to follow children after Fontan are emerging. We hypothesized that significant variability in surveillance testing strategy exists across centers in the United States.

Methods: Eleven academic institutions with established Fontan Care Programs performing a combined, estimated population of 1500-2000 Fontan patients(300 Fontan surgeries/year)were surveyed regarding surveillance testing and basic practice philosophies. Responses were entered into RedCap.

Results: Few practice patterns were identified (table). Neurodevelopmental outcomes were routinely screened in 6 institutions; the rest assessed "as needed". All centers prescribed anticoagulation - most used aspirin alone and none used Coumadin primarily. Only two centers routinely prescribed other medications; one an ACE inhibitor and another a beta-blocker. None routinely used pulmonary vasodilators (including two

Table. Frequency of surveillance testing and consultation (n = 11).

	Not Routinely	Every 6 months	Every Year	Every Two Years > Every 2 years	
	-	-	***Cardiac Testing***	-	-
ECG	0	18%	73%	9%	0
Echo	0	9%	64%	18%	0
Chest x-roy	45%	0	0	9%	45%
Hotter	18%	0	27%	36%	18%
CPET	0	0	9%	36%	55%
Catheterization	36%	0	0	0	64%
Cardiac MRI	9%	0	0	9%	82%
PFT'S	9%	0	9%	9%	73%
	-	-	***Blood Work***	-	-
LFT's/Albumin	0	0	73%	18%	9%
PT/PTT	0	0	64%	27%	9%
CBC	0	0	73%	18%	9%
Iron	18%	0	18%	9%	55%
BNP/NTpro-BNP	9%	0	55%	9%	27%
Alpha-I-AT	9%	0	27%	27%	36%
25-OH Vitamin D	27%	0	36%	0	27%
Electrolytes	0	0	73%	9%	9%
	-	-	***Other Testing***	-	-
Liver U/s	0	0	9%	45%	45%
Liver Biopsy	55%	0	0	0	45%
Urinalysis	36%	0	18%	18%	18%
Cystatin C	27%	0	36%	9%	27%
Urine Prot:Creat	27%	0	27%	9%	36%

	Not Available	Available for Consultation	Available for Consultation (Fontan Expertise)	Sees Routinely	Sees Routinely in an MOC*
Electrophysiology	0	64%	36%	0	0
Heart failure	0	45%	36%	0	18%
GI/Hepatology	0	27%	9%	18%	45%
Pulmonology	0	45%	36%	9%	9%
Endocrinology	0	64%	18%	18%	0
Nephrology	0	73%	18%	0	9%
Hematology	0	91%	9%	0	0
Nutrition	0	45%	18%	18%	18%
Psychology	0	64%	9%	9%	18%

*MOC = Multi-Disciplinary Clinic

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centers at altitude), though when prescribed a PDE-5 inhibitor was most common. Most sites recommended self-limitation of activity and encouraged exercise; 2 allowed participation in competitive athletics; one prescribed specific exercise plans. Eight programs referred adult Fontans to an adult congenital heart disease (ACHD) program while 3 utilized a combination of ACHD and pediatric cardiologists.
Conclusions: Among large academic centers there is a diversity of surveillance strategy after Fontan. However, there are similarities relative to cardiac and secondary organ screening, consultations, and the lack of routine medication use. While most centers proactively involve hepatology, less than half perform liver biopsy routinely. Decreasing practice variability improves quality outcomes. With a rapidly increasing survivial there is urgent need for re-definition of quality outcomes, collaborative practice patterns to decrease variability and innovative research to better understand the complex adaptations of the body to the unique and challenging Fontan physiology.

O2312 - IMPACT OF CONGENITAL HEART SURGERY ON QUALITY OF LIFE IN INFANTS (1 24 MONTHS) A PROSPECTIVE STUDY

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Background: Few studies have prospectively sought to systematically examine the quality of life (QOL) before and after congenital heart surgery. This is especially true of the low resource environments of low and middle-income countries.
Objectives: To determine the impact of heart surgery on the QOL in a cohort of infants with congenital heart disease.
Material and Methods: This was a prospective study from a tertiary care pediatric cardiac center in Southern India. Consecutive infants (1-24 months), admitted for elective congenital heart surgery were included. Validated local language translations of standard QOL infant questionnaires (1-12 and 13-24 months) reported by parents were used (The PedsQL^{TM} Measurement Model for the Pediatric QOL inventory) and 5 standard domains of QOL- physical functioning, physical symptoms, emotional, social, cognitive functioning were tested. Data was collected at preoperative admission and 6 months after surgery.
Results: Over 18-months, 423 infants (1-12 months: 360, 13-24 months: 63) were included. Follow-up data was available for 361 (85.3%) at 6 months. QOL significantly improved from baseline to 6 months after surgery in all domains except cognitive functioning in 1-12 months group; only physical functioning significantly improved in 13-24 months group (see table). Corrective surgery was associated with greater improvements in QOL in all domains except psychosocial health in the 1-12 m group. Lower surgical risk category (RACHS 1-2) was associated with greater improvements in QOL in all the domains in the 1-12 months group.
Conclusion: Congenital heart surgery is associated with improved physical health in all infants while psychosocial health is unchanged in those older than 12 months. Corrective surgery (as against palliative surgery) and lower surgical risk category are associated with better physical and psychosocial improvement.

Table. Change in quality of life from baseline to 6 months in various domains.

	Baseline			6 months		
	Mean	SD	N	Mean	SD	p value*
1-12 Months						
Total Score	81.9	13.3	301	88.1	10.2	<0.001
Physical Health Summary score	80.2	14.9	301	89.1	10.9	<0.001
Physical Functioning	77.3	22.5	295	90.9	13.6	<0..001
Physical symptoms	82.2	13.9	261	88.2	11.4	<0.001
Psychosocial health summary score	83.2	16.0	297	87.3	11.5	<0.001
Emotional funtioning	78.9	19.5	293	82.2	15.1	0.010
Social Functioning	91.4	18.6	261	95.3	12.9	0.002
Cognitive Functioning	91.1	17.8	187	91.8	14.2	0.670
13-24 months						
Total Score	83.1	11.7	54	86.0	11.7	0.180
Physical Health Summary score	79.6	15.6	54	86.2	17.5	0.036
Physical Functioning	77.3	19.6	54	86.1	19.0	0.020
Physical symptoms	81.8	18.0	30	84.9	12.9	0.367
Psychosocial health summary score	85.9	11.0	54	85.6	10.5	0.861
Emotional funtioning	77.3	16.0	54	76.2	15.7	0.693
Social Functioning	95.6	11.8	53	97.6	6.5	0.247
Cognitive Functioning	93.9	12.4	27	88.2	13.6	0.051

N: Number of patients studied; SD: Standard deviation
*Paired t test

O2314 - PRIMARY CONGENITAL ANOMALIES OF CORONARY ARTERIES IN CHILDREN AND ADOLESCENTS

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Background/Objectives: Coronary artery anomalies (CAA) are rare and have the potential to cause present serious myocardial damage, ischemic cardiomyopathy, and sudden cardiac death, but the majority are discovered incidentally. In the present study, children with primary CAA were analyzed to determine the clinical relevance, types of coronary artery anomalies, diagnostic and treatment.
Methods: Retrospective, observational study. Data of our patients below 18 years with CAA in our center until December 2015.
Results: Anomalous coronary arteries were detected in 48 but only in 26 children were isolated (not associated any other cardiovascular disease anomalies or acquired disease). We included 16 anomalous of origin (10 left coronary from pulmonary artery-ALCAPA; 6 anomalous origins from opposite sinus: 5 of right coronary and 1 left coronary); 2 anomalous course; 6 anomalous connections and 2 anomalous number (see Table 1). Mean age at diagnosis was 11,8 years (6m-18y). Murmur was the first symptom, but heart failure was present in 90% of ALCAPA (see Figure). The diagnosis was carried out by echocardiography (ECHO) in 22 cases (84,6%), other imaging modalities (coronary CT, coronary

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angiography or MRI) were used in 15,4% for the diagnosis and to corroborate anatomy in 38% of cases. The treatment was surgical in 11 children (10 ALCAPA, 1 left coronary artery origin from right sinus), 1 interventional catheterization and 4 with medical treatment. After a mean time of follow-up of 4,4 years, one patient died for complications related to diaphragmatic hernia and the rest of patients are stable on clinical observation.
Conclusions: The most frequent anomaly observed in the present study was related to the ALCAPA. The recognition and diagnosis of these anomalies is important and requires specialization in ECHO (main technique for suspicion and diagnosis), coronary angiographic techniques and other imaging modalities.

Table. Summary of primary congenital anomalies of coronary arteries in children

Coronary arteries anomalies	N 26	Group (N)	Clinical main characteristics	Mean age at diagnosis	% ECHO diagnosis	Other diagnostic techniques	Surgical or interventional treatment (%)
Anomalous origins of coronary artery	16	ALCAPA (10)	Cardiogenic Shock/HF (90%)	7,7 m (3 d–3,9 y)	89%	CA (20%)	(100% Surgical)
		Anomalous origins from opposite sinus (6)	Chest pain/ Murmur	9,2 y (1 d-15,6 y)	83%	CCTA (67%)	(1 pt. Surgical)
Anomalous course	2	Inter-arterial course of right coronary artery	Syncope	12 y	100%	CCTA (100%)	-
Anomalous Connections	6	Coronary arteries fistulas (6)	Murmur	2,2 y (1 d-16 y)	89%	MRI (5.5%)/ CA (17%)	Interventional catheterization [1 pt)
Anomalous Numbers	2	Unique coronary artery (2)	Chest pain	4,2 y (1 d–8,5 y)	100%	Stress ECHO (50%)	-

ALCAPA: anomalous left coronary artery from pulmonary artery; CA: coronary angiography; CCTA: coronary CT angiography; ECHO: echocardiography; HF: heart failure

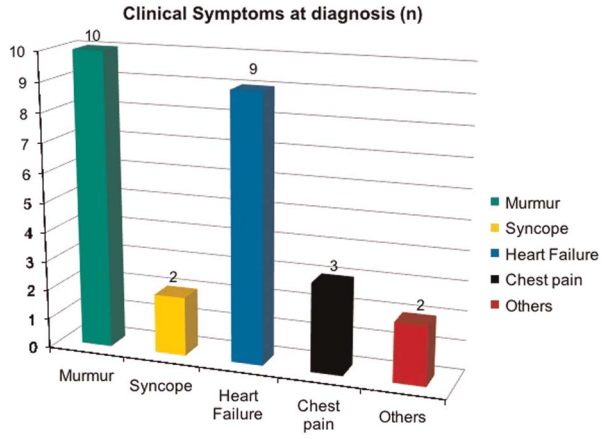

Figure.

O2465 - NEWBORN WELLNESS SCREEN MODIFIED NEWBORN PULSE OXIMETRY SCREENING FOR OUT OF HOSPITAL BIRTHS

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Cardiology Care For Children, Pediatric Cardiology, Lancaster-United States[1]; *Clinic For Special Children, Pediatrics, Lancaster-United States*[2]

Background: Newborn pulse oximetry screening at 24-48 hours of life is recommended to detect critical congenital heart disease (CCHD), but cyanosis can also be due to pulmonary pathology and neonatal infections. Infants born out of hospital often lack a prenatal assessment with ultrasound, have variable rates of maternal GBS testing, and are typically observed only for a few hours after delivery, necessitating a modified pulse oximetry screening protocol.
Methods: Infants had pulse oximetry screening at 1-3 hours of life ("early screen") and again at 24-48 hours ("standard screen") by trained midwives. Midwives were provided training and reliable pulse oximeters (Massimo) to perform the screening. The current AAP-endorsed algorithm for interpreting results was used at both screenings. Midwives followed infants for at least 6 weeks to identify any missed cardiopulmonary pathology or infectious illnesses.
Results: 1212 term infants (50%% male) were screened following parental consent. Prenatal ultrasound was completed for 45% of infants with no CHD identified. 72% of mothers were tested for GBS colonization. Early and standard screens were performed and interpreted correctly for 1077 (89%) infants . At 6 week follow up 1 patient had a small muscular VSD and one was a heterotaxy (unable to obtain saturations); no infections were detected in the cohort.
Conclusion: Preliminary data suggest a combined early and standard newborn pulse oximetry screen can be implemented to detect cyanosis in out of hospital births with a low false positive rate. The early screen is both sensitive and specific with a PPV of 20%. No true positive were identified in the late screen. Of 3 TP 2 had respiratory disease and 1 had complex syndromic cardiopulmonary disease (not CCHD).

Table.

	Early Screen	Late Screen
True positive	3	0
False positive	12	2
True negative	1062	1103
False negative	0	2
Sensitivity	100%	
Specificity	99%	99.8%
NPV	100%	99%
PPV	20%	

O2497 - CORRELATION OF N TERMINAL PRO BRAIN NATRIURETIC PEPTIDE WITH MYOCARDIAL FUNCTION AND ELECTROCARDIOGRAPHIC PARAMETERS IN KAWASAKI DISEASE

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Chul Du Chu De Québec, Université Laval, Pediatrics, Quebec City-Canada[1]; *Boston Children, Pediatric Cardiology, Boston-United States*[2]; *Chu Mère-Enfant Ste-justine, Université De Montréal, Pediatric Cardiology, Montreal-Canada*[3]

Introduction: Pathologic series documented immune myocarditis in nearly all cases of Kawasaki disease (KD), irrespective of coronary complications. N-Terminal Pro-Brain Natriuretic Peptide (NT-proBNP) is useful in the diagnosis of acute KD, but this biomarker has seldom been studied for its relation to myocardial parameters. This study aimed to correlate myocardial response with serum level of NT-proBNP at diagnosis.
Material and Methods: Serum levels of NT-proBNP at diagnosis were retrospectively correlated with echocardiographic

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parameters of myocardial function and electrocardiogram measurements in the acute (1st week), sub-acute (2-3 months) and chronic (6 months to 1 year) phases of KD.

Results: A total of 127 patients with KD were included in this study from 2010 to 2015, among whom 13 (10%) had coronary aneurysms. NT-proBNP (mean 2029 ± 4433 pg/mL) was elevated in 68 patients (61.4%, mean z-score 2.6 ± 1.6, upper quartile 4.67 ± 0.87), with no correlation with the occurrence of coronary aneurysms (p = 0.2). Basic characteristics were comparable among NT-proBNP quartiles. A lower shortening fraction and higher left ventricular mass index (SF z-score -2.8 ± 4.6 and 56.5 ± 13.1 g/m^2, for the upper quartile) were associated with higher NT-proBNP levels (figure 1). Slower mitral deceleration time, lower E/A ratio and higher E/e' ratio also correlated with higher NT-proBNP, reflecting reduced left ventricle compliance. Moreover, higher NT-proBNP was associated with longer QTc duration, wider QT dispersion and longer PR interval (figure 2). The correlations with elevated NT-proBNP persisted at 3 months but disappeared at 1 year follow-up, with the exception of QTc which maintained a trend towards longer duration.

Conclusion: Acutely elevated NT-proBNP in KD is associated with systolic, diastolic and electrical disturbances, during the acute and sub-acute phases of the disease. These findings suggest myocardial involvement in KD, even in the absence of coronary aneurysm. Therefore, long-term follow-up in patients with elevated NT-proBNP at diagnosis should be seriously considered.

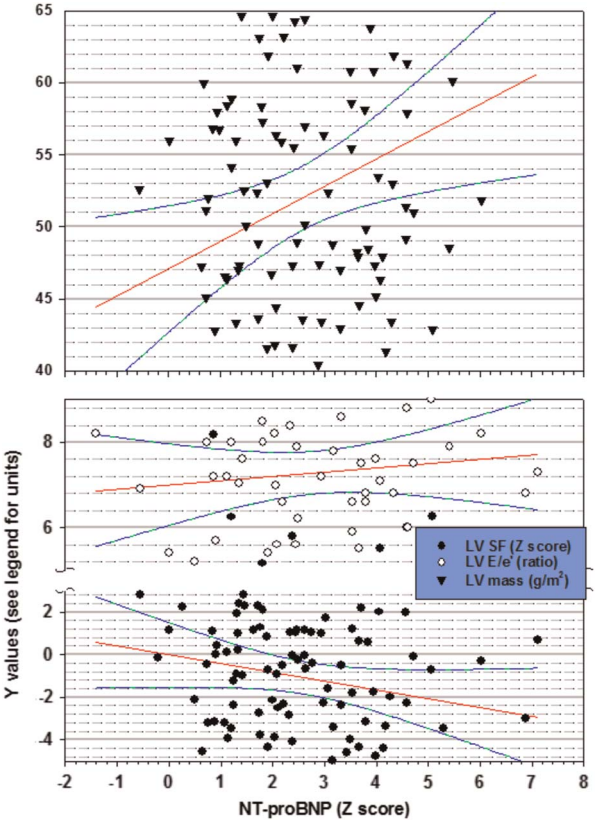

Figure 1.
Linear Regression of N-Terminal Pro-Brain Natriuretic Peptide Z-score Correlated to Shortening Fraction Z-Score (decrescendo), Left Ventricular Mass Index (crescendo) and E/e' Wave (crescendo).

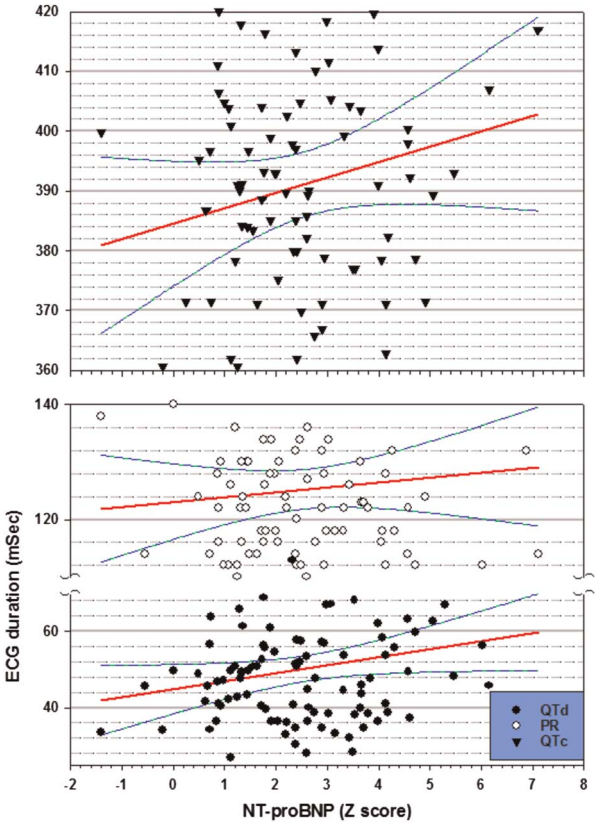

Figure 2.
Linear Regression of N-Terminal Pro-Brain Natriuretic Peptide Z-Score Correlated to Corrected QT interval, QT dispersion and PR interval (all crescendo).

O2657 - RELATIONSHIP OF AMBULATORY BLOOD PRESSURE WITH LEFT VENTRICULAR GLOBAL LONGITUDINAL STRAIN

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Background: Left ventricular (LV) global longitudinal strain (GLS) is an early indicator of subclinical cardiac dysfunction, even when LV ejection fraction (LVEF) is normal, and is an independent predictor of cardiovascular events. Ambulatory blood pressure (BP) is a good predictor of cardiovascular events, including heart failure. We investigated the association of ambulatory BP measurements with subclinical LV systolic dysfunction in a paediatric community-based cohort with normal LVEF.

Methods: Two-dimensional speckle-tracking echocardiography and 24-hour ambulatory BP monitoring were performed in 49 children (mean age 11 ± 2 years; 56% men) with LVEF ≥55%. All children were sent for cardiac evaluation because of 3-high-BP-values (> p97 according to height) were reported by their paediatricians. Univariable and multivariable linear regression analyses were used to assess the associations of BP measures with GLS.

Results: Higher ambulatory BP values were consistently associated with impaired GLS. After adjustment for pertinent covariates (age, sex, race/ethnicity, body mass index), ambulatory systolic and diastolic BPs was independently associated with GLS (P ≤0.001 for all ambulatory BPs).

Conclusions: Ambulatory BP values are significantly associated with impaired GLS. Office BP is a strong surrogate for ambulatory BP

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and GLS. Ambulatory BP monitoring might have a role in the risk stratification of hypertensive children for early LV dysfunction.

O2725 - CARDIAC DISEASE IN PEDIATRIC PATIENTS PRESENTING TO CARDIOLOGY DEPARTMENT WITH CHEST PAIN

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Background: Chest pain in children and adolescents is one of the most common complaint for referral to pediatric cardiologists. In this study, it is aimed to determine the proportion of children referred to pediatric cardiology clinic for chest pain diagnosed with a cardiac cause of chest pain.
Methods: We reviewed 635 (325 male, 310 female and 6-18 years) patients who presented with a chief complaint of chest pain and no history of cardiovascular disease prospectively. Eligible patients were evaluated with regard to demographic and chest pain characteristics, past medical history, family history, findings on physical examination, electrocardiogram and chest radiography. If necessary, futher cardiac evaluation were performed.
Results: 635 patients reported with chest pain during the study period. Only 24 (3.7%) of all patients were determined to have chest pain of cardiac origin. Those with cardiac- related chest pain had been consisted of 10 myopericarditis, 10 mitral valve prolapse, 2 myocardial bridge, 1 coroner vasospasm and 1 hypertrofic cardiomyopathy. All patients had an electrocardiogram. Thirty-eight percent of the cardiac related chest pain had abnormal electrocardiogram compared to those without cardiac related chest pain at 4.3% ($p < 0.001$). Furthermore, when compared these groups, for high risk chest pain characteristics, medical or family history and abnormal cardiovascular exam findings, in cardiac group, these pathologies were significantly high ($p < 0.001$).
Conclusion: Cardiac related chest pain is rare but potentially serious. Although a pediatric cardiology referral may provide reassurance to the primary care and emergency physicians, our results reveal that cardiac pathology for pediatric chest pain are rare. An electrocardiogram in addition to detailed history and careful physical examination is appropriate approach to optimize testing and referral patterns while maintaining a high detection rate for relatively uncommon but significant cardiac disease.

O2999 - CHILDREN WITH CONGENITAL HEART DISEASE (CHD) ARE THEY SAFE TO FLY

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Royal Brompton Hospital, Paediatric Cardiology, London-United Kingdom[1]; Royal Brompton Hospital, Respiratory Physiology, London-United Kingdom[2]; Royal Brompton Hospital, Paediatrics, London-United Kingdom[3]

Background: Healthy children de-saturate by approximately 4% during air flight as cabin pressurisation (5-8000 feet equivalent) results in a reduced fraction of inspired oxygen to the equivalent of 0.15. Little is known about the effect of in flight hypoxia on children with CHD.
Aim: To measure oxygen saturations of children with CHD during a hypoxic challenge /'fitness to fly' test to guide recommendations for supplementary oxygen.
Method: Observational study including children with CHD between 0-≤16 years. Exclusion criteria were pulmonary hypertension; pre-existing oxygen requirement; significant respiratory disease; and baseline SpO2 of ≤74%. A wide spectrum of cardiac diagnoses, including post-operative children, were studied. During testing the child sits inside a plethysmograph (body box) in which oxygen level is reduced to 15% over 5 minutes. Supplemental oxygen was given if saturations fell by more than 15% or to below 70% indicating test failure.
Results: See table & graphs – Figure 1 (69 participants).
Conclusions: Children with CHD having baseline cyanosis (75-94%) or normal saturations (95-100%) with potential for right to left (Rt-Lt) shunting, may desaturate by >15% during hypoxic challenge flight simulation. Hence we recommend children with CHD, with either cyanosis or potential for Rt-Lt shunting, undergo a fitness to fly simulation test prior to real flight, to identify those who should be given oxygen during flight to prevent significant desaturation.

Groups	Baseline SpO_2% (median)	Lowest SpO_2% (median)	Number of Participants Failing Test (%)
Group A (n=37)	95 - 100 (98.3%)	84 – 95 (89.8%)	4 (11%)
Group B (n=32)	75 - 94 (84.8%)	65 - 84 (76.6%)	5 (16%)

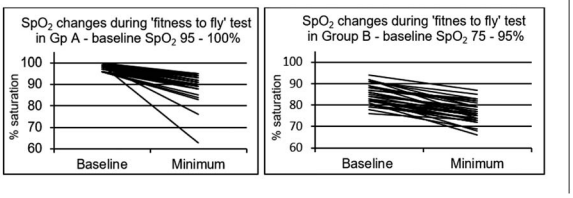

Figure.

ANESTHESIA

O1110 - PREOPERATIVE NUTRITIONAL DEFICITS WORSEN OUTCOMES FOLLOWING PEDIATRIC CARDIAC SURGERY

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Seattle Children, Anesthesiology and Pain Medicine, Seattle-United States[1]; Seattle Children, Critical Care, Seattle-United States[2]

Introduction: Children with congenital heart disease are at increased risk of acute and chronic malnutrition due to increased metabolic demands and diminished energy intake. In this study, we evaluated the impact of nutritional status on outcomes after pediatric cardiac surgery in a large cohort of children.
Methods: We conducted a retrospective analysis of patients from age 0 to 5 years undergoing cardiac surgery at Seattle Children's Hospital from 2006 to 2015. Anthropometric indices, including height-for-age, weight-for-age, and weight-for-height z-scores (HAZ, WAZ, WHZ), were calculated for each patient using WHO standards. We used multivariate regression modeling controlling for age and surgical complexity to examine the impact of nutritional status on surgical outcomes including 30-day mortality, cardiac arrest, infection, duration of mechanical ventilation, ICU LOS, and hospital LOS.
Results: Nutritional deficits, indicated by low HAZ and low WAZ were associated with increased mortality. This relationship was nonlinear; nutritional status has little impact on mortality until deficits become severe with HAZ or WAZ ≤-2. In this range, each additional unit decrease in HAZ or WAZ is associated with a 2.87% or 2.14% increased risk of mortality respectively. Low HAZ and WAZ were also associated with increased risk of cardiac arrest, infection and longer mechanical ventilation and ICU and hospital LOS.
Conclusion: Nutritional deficits increase the risk of mortality and other adverse outcomes after pediatric cardiac surgery. From our data, HAZ has the strongest relationship with mortality; thus,

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HAZ should serve as a useful screening index for patients at risk for complications related to malnutrition. The dramatic increase in mortality with HAZ and WAZ ≤ -2 indicates that nutritional optimization efforts should be focused on this population. Given the association between nutritional deficits and longer mechanical ventilation and hospital and ICU LOS, focused nutritional optimization has the potential for significant cost savings.

Table. Risk of outcomes based on nutritional status

	30-day Mortality	Cardiac Arrest	Infection
HAZ	Nonlinear*	-0.012* (-0.018,-0.006)	-0.011* (-0.017,-0.004)
HAZ <= -2	-0.029* (-0.050, -0.007)		
WAZ	Nonlinear*	−0.007* (-0.013, -0.002)	-0.008* (-0.014, -0.002)
WAZ <= -2	-0.021* (-0.042, -0.001)		
WHZ	-0.003 (-0.008, 0.002)	0.004 (-0.002, 0.010)	0.002 (-0.006, 0.010)
	Mech. Vent. (days)	ICU LOS (days)	Hospital LOS (days)
HAZ	-0.07* (-0.10, -0.04)	-0.25* (-0.34, -0.15)	-0.57* (-0.74, -0.39)
WAZ	-0.08* (-0.11, -0.05)	-0.22* (-0.33, -0.12)	Nonlinear*
WHZ	Nonlinear*	-0.01 (-0.11, 0.10)	Nonlinear*

For categorical variables, each outcome probability is reported based on a unit change in the z score. For non-categorical variables, the value in the table represents B from robust linear regression model, or the coefficient of the linear relationship between the independent and dependent variables. The values in parentheses represent 95% confidence intervals. *Statistically significant results are starred.

O1863 - OUTCOMES OF CHILDREN WITH CONGENITAL HEART DISEASE UNDERGOING BYPASS SURGERY BY ROUTE OF TRACHEAL INTUBATION IN THE UNITED STATES AN ANALYSIS OF THE SOCIETY OF THORACIC SURGEONS CONGENITAL HEART SURGERY DATABASE

Nathaniel Greene[1], Dylan Thibault[2], Eileen Raynor[3], Alice Wang [4], David Vener [5], Roland Matsouaka[2], Marshall Jacobs[6], Jeffrey Jacobs[7], Kevin Hill[8], Warwick Ames[1]

Duke University, Anesthesiology, Durham, Nc-United States[1]; Duke University, Biostatistics, Durham, Nc-United States[2]; Duke University, Otolaryngology, Durham, Nc-United States[3]; Duke University, Surgery, Durham, Nc-United States[4]; Baylor College of Medicine, Anesthesiology, Houston, Tx-United States[5]; Johns Hopkins University, Surgery, Baltimore, Md-United States[6]; Johns Hopkins All Children, Surgery, Saint Petersburg, Fl-United States[7]; Duke University, Pediatric Cardiology, Durham, Nc-United States[8]

Introduction: In children undergoing cardiopulmonary bypass (CPB) surgery, nasal intubation is more common than in adults, and sometimes preferred due to perceived benefits of less post-operative sedation and a lower risk for accidental extubation. This study sought to describe the practice of nasal intubation in the pediatric population undergoing CPB surgery and assess the risks/benefits of a nasal route against an oral one.
Methods: Patients <18 years of age in the STS Congenital Heart Surgery Database (01/2010–12/2015) were included. Multivariable modeling was used to assess the association between route of tracheal intubation and a composite measure of infection risk. Covariates were included to adjust for important patient characteristics, case complexity, and center effects. Secondary outcomes included length of intubation, hospital length of stay, and airway complications including accidental extubations.
Results: Unadjusted analysis of the entire cohort of 26,788 operations from 50 centers showed nasal intubation to be associated with a higher composite infection rate ($p = 0.004$), longer intubation length ($p < 0.0001$), longer post-operative length of stay ($p < 0.0001$), and a similar rate of accidental extubation ($p = 0.82$). Multivariable analysis in infants and neonates showed the nasal route of intubation was not associated with the infection composite, but was associated with a shorter length of stay, and a shorter length of intubation. Restricting to centers with at least 100 cases per year showed a significant interaction between age and intubation route with a risk change occurring at approximately 6 months of age ($p = 0.003$, Figure 1).
Discussion: While older children undergoing nasal intubation trend similar to the adult population with an increased risk of infection, nasal intubation in children <6 months may confer some protection. This may be related to the timing of maxillary/ethmoid sinus development and/or development of adenoid tissue, acting as a source of bacteria for nasal endotracheal tubes.

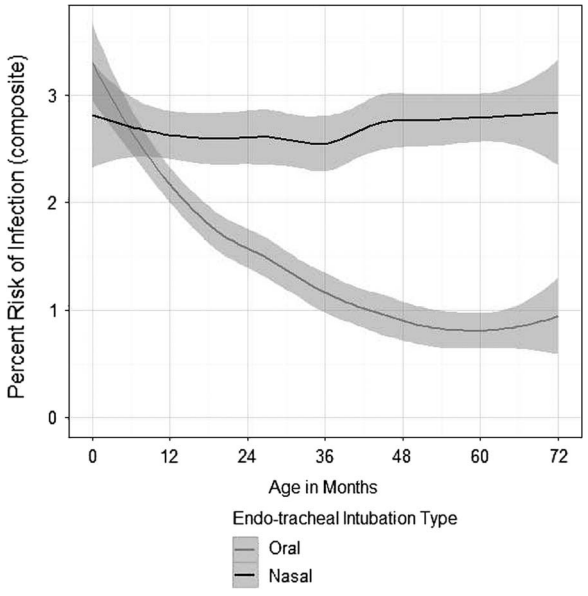

Figure 1.
Risk of Infection for Oral vs Nasal Intubation by Age (72 months and under) with Smoothed line.

O1999 - BASELINE MA FF FLEV AND FIBRINOGEN LEVELS IN NEONATES INFANTS AND CHILDREN WITH CONGENITAL HEART DEFECTS

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Emory University School of Medicine, Anesthesiology, Atlanta-United States[1]; Emory University School of Medicine, Pathology, Atlanta-United States[2]

Introduction: In children undergoing cardiac surgery, fibrinogen restoration is important in correcting coagulopathies after cardiopulmonary bypass (CPB) (1). However, determining the clinically appropriate measure of fibrinogen for this correction is less than clear. Clauss-method measurement of fibrinogen levels is time consuming and is performed on plasma samples activated with excess thrombin. Thromboelastographic (TEG®; Haemonetics) measurement of "functional fibrinogen" is quicker and is performed on whole blood (WB) samples using a GpIIb/IIIa receptor blocker to inhibit platelets and thus isolate the contribution of fibrinogen to clot strength. Haemonetics has developed a proprietary measure of this functional fibrinogen ("FLEV") derived

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from the maximum amplitude of these modified TEGs (MA-FF). However, MA-FF and FLEV reference ranges have not been determined for young children. The primary goal of this study was to establish these ranges. A secondary goal was to seek correlation between MA-FF or FLEV values and fibrinogen levels measured by the standard laboratory Clauss method.
Methods: In this prospective observational study, we selected 15 consecutive patients from 5 age groups (<30 days, 1-3 months, 3-6 months, 6-12 months, 12-24 months) and 30 patients >24 months. Patients with bleeding issues or on drugs affecting coagulation were excluded. Samples were collected after induction of anesthesia and prior to surgical incision. MA-FF, FLEV, Clauss derived fibrinogen levels, and platelet counts were measured by our hospital laboratory and values were compared among age groups using ANOVA with post-hoc Tukey-Kramer analyses. Correlation between MA-FF or FLEV and fibrinogen levels was sought using Spearman's correlation with Bonferroni correction.
Results: Results are displayed in the Table. Statistically higher MA-FF and FLEV values were found in infants 1-3 months compared to children 12-24 or >24 months. There were no differences among fibrinogen levels in any of the age groups. Platelet counts were higher in infants 1-3 months compared to children >24 months. MA-FF and FLEV values correlated with Clauss measured fibrinogen levels only after 6 months ($r \geq 0.7$; $p < 0.001$ in all groups >6 months).

Table 1. Comparison of TEG values and laboratory tests.

	MAFF	FLEV (TEG)	Fibrinogen Level (Clauss)	Platelet count	Hematocrit
<30 days	22.2 ± 5.3	406 ± 97	258 ± 82	328 ± 81	43.6 ± 8.2^
1-3 months	26.3 ± 5.7⋆	479 ± 104⋆	283 ± 70	395 ± 103#	37.2 ± 7.1
3-6 months	24.0 ± 3.8	437 ± 70	258 ± 65	336 ± 113	36.3 ± 6.2
6-12 months	22.1 ± 4.3	404 ± 78	271 ± 67	328 ± 150	36.9 ± 5.4
12-24 months	19.8 ± 5.8	361 ± 106	255 ± 76	321 ± 92	39.0 ± 5.2
>24 months	21.2 ± 4.0	378 ± 79	268 ± 53	278 ± 65	39.2 ± 5.0

⋆p <0.05 versus 12-24 months and >24 months for MA-FF and FLEV values.
#p <0.05 versus >24 months for Platelet Count.
^p <0.05 versus 3-6 months and 6-12 months for Hematocrit.

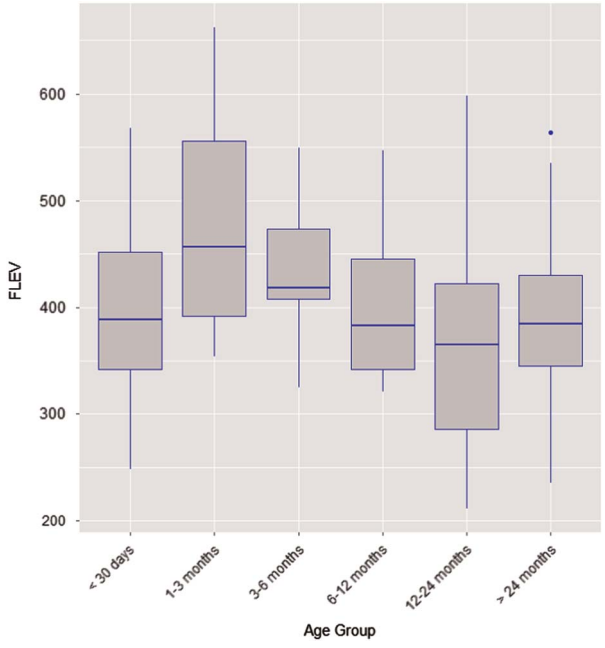

Figure 1.
FLEV values per age Group.

Discussion: This preliminary study establishes reference ranges for MA-FF and FLEV values in 6 age groups of children. These ranges are necessary to guide TEG-directed fibrinogen replacement after CPB. In neonates and infants <6 months WB-derived MA-FF or FLEV values were higher and did not correlate with plasma-derived Clauss fibrinogen levels. This may reflect TEG's ability to assess collective interactions of multiple pro- and anti-coagulant deficiencies (2) or molecular and structural differences in fibrinogen observed in these young infants (3). TEG values may be more useful when making clinical decision in young infants.

1. Anesth Analg 1997;85:1196.
2. Am J Ped Hemat Oncol 1990;12:95.
3. Anesthesiology 2016;124:1021.

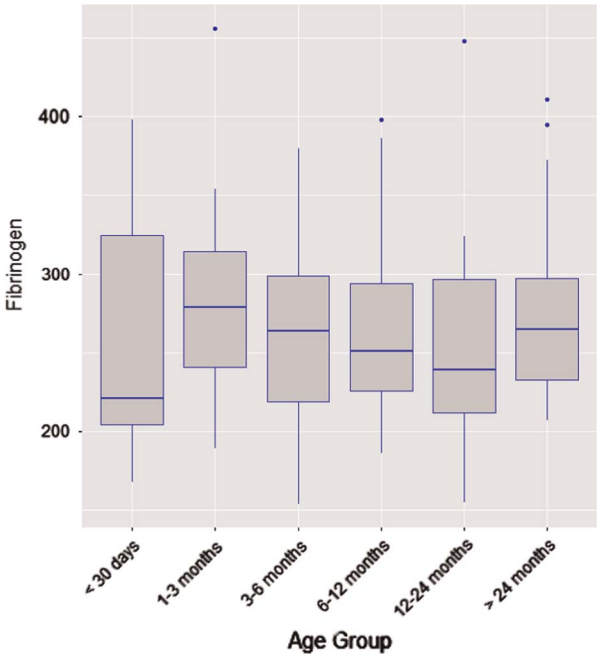

Figure 2.
Fibrinogen Levels per age Group.

O2207 - ANESTHETIC MANAGEMENT OF ABO INCOMPATIBLE PEDIATRIC ORTHOTOPIC HEART TRANSPLANT A CASE SERIES

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Background: Mounting data have accumulated demonstrating equivalent outcomes and reduced waiting list mortality for pediatric patients receiving ABO-incompatible (ABOi) orthotopic heart transplant (OHT). We present a case series of patients undergoing ABOi OHT and discuss the unique perioperative challenges that arise, including a previously undescribed hemodynamic pattern observed.
Case Series Presentation: There were a total of seven patients undergoing ABOi OHT at our institution from October 2011 to October 2016. Average age was 6.7 months. Five patients' etiology of heart failure was idiopathic dilated cardiomyopathy (DCM), one was due to familial DCM and one was due to hypertrophic CM. Five were on inotropes or vasopressors

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preoperatively. Five received 3x blood volume exchange transfusion (ET), one received 5x and one received 1x blood volume ET. All were supported with epinephrine and milrinone infusions post-CPB. Of note, during the post-CPB period four patients required near maximal milrinone at 0.8-1 mcg/kg/min, two patients required nitroglycerine boluses, and one required vasodilator infusion for hypertensive urgency.

Perioperative Management: At our institution, ET is undertaken according to a protocol based on patient and donor ABO group, as well as isohemagglutinin titer levels. The CPB circuit is primed with recipient-compatible pRBCs and donor-compatible plasma and platelets. Our protocol entails 3x blood volume ET if isohemagglutinin titers are less than 1:2, and 5x blood volume ET if titers are greater than 1:2. ET produces vasodilatory and hypovolemic hypotension lasting about 3-5 minutes. This can be a challenge in patients with already-failing hearts, thus anticipatory management and close communication with perfusionists is critical. Separation from CPB and the post-CPB period during OHT is typically characterized by vasoplegia, as well as donor organ dysfunction due to ischemia and denervation. However, in our cohort several patients demonstrated unexpected clinically significant hypertension during the immediate post-CPB period requiring antihypertensive treatment.

O2401 - DOUBLE BLIND RANDOMIZED PLACEBO CONTROLLED TRIAL COMPARING THE EFFECTS OF ANTI THROMBIN VS PLACEBO ON THE COAGULATION SYSTEM IN INFANTS WITH ANTI THROMBIN DEFICIENCY UNDERGOING CONGENITAL CARDIAC SURGERY

Rebecca Scholl[1], Claudia Benkwitz[2], Manuel L. Fontes[3], Nirmish Shah[4], Robert D.B. Jaquiss [5], Andrew J. Lodge [5], Warwick A. Ames[1], Mayumi Homi[1], Kelly A. Machovec[1], Edmund H. Jooste[1]

Duke University Hospital, Pediatric Anesthesiology, Durham-United States[1]; Vanderbilt University, Pediatric Anesthesiology, Nashville-United States[2]; Yale, Pediatric Anesthesiology, New Haven-United States[3]; Duke University Hospital, Medicine, Durham-United States[4]; Duke University Hospital, Congenital Cardiac Surgery, Durham-United States [5]

Introduction: Anti-thrombin (AT) is crucial for heparin to be effective and for the prevention of thrombosis. We tested the hypothesis that pre cardiopulmonary bypass (CPB) normalization of AT levels to 100% would improve anticoagulation during CPB, attenuate activation of the coagulation cascade, as measured by fibrin degradation products- D-Dimers, and mitigate microthrombosis in the post-operative period.

Methods: This is a randomized, double blinded, placebo controlled study in infants with AT levels less than 70% undergoing CPB at 2 academic centers. Half were given AT (Thrombate®) to normalize to 100% functional assay and the other half received placebo. We recorded demographics, laboratory values, hemostatic agents, blood products administered, operating room times, 24-hour chest tube output, and occurrence of post-operative thrombosis. Summary statistics, pooled T-test, Fisher exact test and Wilcoxon Rank-Sum test were performed.

Results: Over 24 month period 223 patients were screened, 68 met inclusion criteria, 40 were randomized, and 1 (in Placebo group) was withdrawn during the study due to ECMO initiation in the operating room. Thirty percent of screened patients had AT levels <70%. Table 1 shows demographic and peri-operative variables. Notable differences were observed between the two groups including higher first post-heparin activated clotting time (ACT) and AT levels in the treatment group. Table 2 shows Laboratory results. Significantly, 24 hours post-operative chest tube output, overall blood product transfusions and D-Dimer production were all lower in the AT group. No difference in major adverse events was noted.

Conclusion: Replacement of AT improves anticoagulation during CPB without increased rates of bleeding or adverse events. Furthermore our results suggest extension of these beneficial effects into the postoperative period, reflected by significantly less postoperative bleeding and transfusions, and generation of D-Dimers. We conclude that treatment of low AT levels with exogenous AT is helpful in infants undergoing cardiac surgery.

Table 1. Demographics and perioperative variables.

	AT Group (n = 20)	Placebo group (n = 19)	p value
Age (days)	9.8 ± 19.7	17.2 ± 30.2	0.174
Gender (M:F)	14:06	13:06	0.915
Weight (kg)	3.4 ± 0.5	3.5 ± 0.6	0.792
Aristotle score level			0.304
Level 2 (includes Aristotle scores 6.0-7.9)	*3 (15.0%)*	*1 (5.3%)*	
Level 3 (includes Aristotle scores 8.0-9.9)	*7 (35.0%)*	*4 (21.1%)*	
Level 4 (includes Aristotle scores 10.0-15.0)	*10 (50.0%)*	*14 (61.5%)*	
ACT baseline (sec)	129 ±22	120 ± 13	0.141
Pre CPB Heparin dose determined by HMS machine (U/kg)	532 ± 64	602 ± 154	0.106
ACT post heparin (sec)	**740 ± 221**	**583 ± 127**	**0.041**
Heparin dosing - Total on CPB (U)	**4372 ± 1627**	**5520 ± 1924**	**0.055**
Total CPB time (min)	185 ± 60	212 ± 66	0.186
Total Cross clamp time (min)	87.4 ± 54	101.4 ± 32	0.329
Protamine dose determined by HMS machine (mg/kg)	11 ± 7.6	9.3 ± 2.8	0.545
Protamine to chest closure time (min)	109.8 ± 56.5	105.1 ± 59	0.757
Chest Tube output (protamine time plus 24 hrs) (mls)	**74.9 ± 50**	**138 ± 94**	**0.016**
Exposure to any Blood products in 24 h Postop (n)	6	11	
Total 24 h Postop Blood product unit exposures (n)	**6**	**19**	**0.000003**
24 h Postop FFP Unit exposures (n)	*1*	*3*	
24 h Postop Platelet Unit exposures (n)	*0*	*4*	
24 h Postop Cryo-precipitate Unit exposures (n)	*0*	*3*	
24 h Postop Red Blood cell Unit exposures (n)	*5*	*9*	
Major Adverse Events	8	8	0.894

Results represented by Mean ± SD. HMS machine - Hemostasis Management System machine; CPB - Cardiopulmonary bypass; AT- Anti-Thrombin; Postop - Postoperative.

Table 2. Study lab Values.

	Time Point	ATIII Group (n = 20)	Placebo group (n = 19)	p value
ATIII functional assay (%)	T1 (Baseline)	54 ± 12	54 ± 13	0.982
ATIII functional assay (%)	**T2 (30 min after study drug)**	**99 ± 19**	**49 ± 16**	**<0.0001**
ATIII functional assay (%)	**T3 (30 min on CPB)**	**83 ± 20**	**55 ± 27**	**0.0007**
ATIII functional assay (%)	**T4 (just prior to coming of CPB)**	**87 ± 34**	**63 ± 28**	**0.048**
ATIII functional assay (%)	**T5 (Arrival in ICU)**	**82 ± 18**	**63 ± 19**	**0.003**
ATIII functional assay (%)	T6 (POD 2)	58 ± 15	57 ± 14	0.84
ATIII functional assay (%)	T7 (POD 4)	70.7 ± 20	66 ± 17	0.475
D Dimer (mcg/ml)	T1 (Baseline)	1.2 ± 0.8	1.3 ± 0.8	0.855
D Dimer (mcg/ml)	T5 (Arrival in ICU)	1.0 ± 0.9	1.3 ± 1	0.225
D Dimer (mcg/ml)	T6 (POD 2)	1.5 ± 1	2.3 ± 2	0.313
D Dimer (mcg/ml)	**T7 (POD 4)**	**3.6 ± 1.7**	**6.8 ± 4.2**	**0.008**

Results represented by Mean ± SD. CPB - Cardiopulmonary bypass, ATIII- Anti-Thrombin III.

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ELECTROPHYSIOLOGY

O1105 - PERMANENT CARDIAC PACING IN CHILDREN IN A SUB-SAHARAN TERTIAIRY CENTRE

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Background: Pacing in children is mainly performed in the setting of congenital or post-surgical complete heart block and less frequently in some surgical patients with sinus node dysfunction. Few data are available concerning Africa. The goal of the study was to investigate the indication, prevalence of permanent cardiac pacing in children and follow -up in St. Elizabeth Catholic General Hospital Shisong, Cardiac centre.
Patients and Methods: From the 10th November 2010 till 10th November 2016, out of 156 patients who underwent a device implantation in the Cardiac Centre Shisong, 9 (5,7%) were children. Data extracted from the records of implanted patients were demographics, clinical indications for device implantation, the electrocardiogram, the echocardiogram.
Results: In the Cardiac Centre Shisong, for bradypacing were implanted 156 pace makers. 10 patients having atrial fibrillation with low ventricular response benefited from a single chambered pace maker. Sick sinus node was diagnosed in 46 patients; 40 cases benefited from a dual chambered pace maker and 6 from a single chambered pace maker. 100 cases had complete atrio-ventricular block and were implanted 5 – patients single chambered and 95 dual chambered pace makers. The pediatric population consisted of 9 children, 7 teenagers suffering from sick sinus node with syncopal attacks, and two children with post surgical complete atrioventricular block in whom was surgically corrected a ventricular septal defect. All patients were above 15 kg and benefited from endovenous pacing, the conventional approach with left subclavian vein puncture being the one used. No complications were seen postoperatively and during the longterm follow-up was registered a death due to severe malaria.
Conclusion: In the Cardiac centre Shisong, permanent pacing in adult and children is of actuality with good results. The main indications are sick sinus node and complete postsurgical block.

O1126 - ALIVECOR FOR LIFE A DIFFERENT APPROACH TO CARDIAC EVENT MONITORING IN CHILDREN

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Background: The AliveCor heart monitor (Kardia) attaches to a mobile device running the Alive ECG app and has proved successful in diagnosing arrhythmias in adults but there are little data of its use in children. The aim of our study was to evaluate the effectiveness of issuing an AliveCor for Life in children with palpitations.
Methods: Children were eligible for the AliveCor monitor if they were >2 years old, had symptoms of palpitations less frequently than once a week and if they or their parents had a device compatible with the AliveCor monitor. An NHS email account was set up exclusively for receiving ECG recordings emailed via the AliveCor App. The email account was checked twice daily by an experienced cardiac physiologist. The children were given the AliveCor monitor to keep. The number of ECG recordings received, number of arrhythmias documented, quality of ECG recordings, annual costs and patient satisfaction were evaluated.
Results: AliveCor monitors were issued to 50 patients, aged 4 years to 17 years (mean 11 years). 565 ECG recordings were obtained from the AliveCor monitors of which 80 showed arrhythmia (14.2% yield). 523(93%)of the ECG recordings were considered acceptable for analysis and 42 uninterpretable. The AliveCor monitors cost £4,000 for 50. There were no maintenance or retrieval costs and in three patients the AliveCor for Life gave an alternative to an implantable loop recorder (saving a cost of £6,000 for 3 implants) There was high patient satisfaction especially in the teenage group. On comparison with our established methods for non invasive cardiac event monitoring for palpitations, the AliveCor outperformed with respect to ECG recordings, diagnostic yield, annual costs and patient satisfaction.
Conclusions: An AliveCor for Life allows effective, simple and affordable longterm ECG event monitoring in children that is highly acceptable to the patient and parent.

O1194 - VENTRICULAR TACHYCARDIA IN PEDIATRIC POPULATION

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Objective: To delineate the outcome for ventricular tachycardia (VT) in pediatric population.
Methods: From 1991 to 2015, patients who developed sustained VT at the age of 0 to 18 years in a referral center were enrolled.
Results: A total of 116 patients (67 male/49 female) had documented VT, and 62 (53.4%) patients had associated heart disease, namely cardiomyopathy in 21 (18.1%), structural heart disease in 23 (19.8%), channelopathy in 14 (12.1%), and arrhythmogenic right ventricular cardiomyopathy in 6 (5.2%), and some of them with two types of associated heart disease. Idiopathic VT (65 patients) was the most common types of VT; 72.6% with right bundle branch block (RBBB) morphology and 27.4% with left bundle branch block morphology. Forty-one patients received catheter ablation with a success rate of 90.4%, a recurrence rate of 14.3%. None of the patients died during 5.8 + -5.9 years follow-up. Cardiomyopathy with VT exhibited the highest mortality rate (47.6%), especially for those with hypertrophic and restrictive cardiomyopathy. Among 16 patients with coexisting VT and dilated cardiomyopathy, 7 exhibited improved heart function after VT controlled, and could be predicted by onset symptoms, monomorphic QRS morphology, and duration between diagnosis of cardiomyopathy and VT. VT associated with structural heart disesase carries a high risk of mortality, but this risk has decreased after aggressive intervention in recent 15 years. VT with channelopathy can often be controlled by medication , but those with fetal onset has specially high mortality rate and aggressive intervention may be necessary in these patients.
Conclusions: VT in pediatric population, although may carry high mortality when associated with structural anomaly or cardiomyopathy, often has favorable outcome after prompt intervention. The course and clinical outcome is highly dependent on its etiology and age of onset.

O1553 - LEFT VENTRICULAR APICAL PACING IN CHILDREN – FEASIBILITY AND LONG–TERM EFFECT ON VENTRICULAR FUNCTION

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Background: Left ventricular (LV) pacing has been reported to preserve LV function in chronically paced children with atrioventricular (AV) block. We sought to evaluate long-term results of LV apical pacing (LVAP) in patients with both structurally normal heart and congenital heart disease.
Patients and methods: 36 patients with complete spontaneous (N = 22, group A) and surgical AV block (N = 14, group B, systemic LV in all) received an epicardial VVIR (N = 19) or DDDR (N = 17) pacemaker at the median age of 1.69 (IQR 0.04-4.39) years. Bipolar ventricular pacing leads (Medtronic 4968) were placed at the LV apex using a subxiphoid approach (N = 18) or sternotomy (N = 18). After median followup of 2.9 (IQR 1.9-6.2) years echocardiography and exercise stress testing was performed. Data were compared to age-matched normal controls (N = 25, group C).
Results: Pacemaker implantation was uneventful, there was no death and all patients were on LVAP at the end of follow-up. Probability (3 and 6 years after implantation) of absence of pacemaker-related surgical revision (elective generator replacement excluded) was 89.0 and 89.0%, resp. Probability of freedom from battery depletion was 91.4% and 77.8%, resp. Ventricular thresholds at given pulse duration did not change between discharge and last follow-up. There was no significant difference in LV end-diastolic volume index (LVEDVi) and ejection fraction (LVEF) between groups A, B and C at last follow-up. Interventricular and intra-LV synchrony was preserved in groups A and B. Maximum oxygen uptake was, however, lower in group A = mean 33.5 (SD 5.8) and B = 33.9 (6.1) ml/kg/min as compared to group C = 40.8 (6.6) ml/kg/min, p = 0.009.
Conclusions: LVAP carries complete preservation of LV function and synchrony. Pacing-related complications are rare and probability of continued LVAP is excellent despite patient growth.
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O1783 - A NEW HOLTER TECHNIQUE TO DIAGNOSE AND RISK STRATIFY CHILDREN WITH LONG QT SYNDROME

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Background/Hypothesis: Diagnosing long QT syndrome (LQTS) and predicting risk of cardiac events currently relies on the manifestation of a prolonged QTc or life-threatening event. At least 30% of LQTS subjects have a normal QTc on 12-lead ECG. We hypothesized assessment of repolarisation duration from a single Holter recording may identify disease status and correlate with risk of cardiac events.
Materials and Methods: 145 Holter recordings were analysed. LifeCard CF monitors and DelMar Impressario software were used to calculate mean-QTc and mean-RTPc (R-wave-peak T-wave). Only heart-rates 40-120BPM were included. A single experienced cardiac technician used the semi-automated measurement technique. End-QT was defined by zero crossing point. Bazett heart-rate correction was used. Normative data were taken from 56 controls (0-16years). The highest mean control value was taken as the upper limit of normal (ULN; QTc = 450ms, RTPc = 312ms). Of 89 children assessed for possible LQTS (0-16years); 68 ulitmately were diagnosed with LQTS (44 KCNQ1-positive, 18 KCNH2-positive, 6 gene-unknown (Schwartz scores 4)) and 21 were cascade gene-negative relatives. Results were evaluated against lifetime cardiac events over 987patient-years.
Results: Mean-QTc (48339ms) and mean-RTPc (34534ms) were longer in LQTS individuals than gene-negative relatives and controls combined (Mean QTc 427 + /-20ms;mean RTPc 291 + /-18ms $p < 0.0001$). The mean-RTPc was the same in Gene-negative children and controls, but mean-QTc was 13ms longer ($p = 0.008$). All children with CA (n = 4) were ≥20ms over ULN. 17/18 syncopal patients were ≥5ms above QTc-ULN or ≥15ms above RTPc-ULN. RTPc specificity was superior to QTc. Sensitivity/Specificity (%) of mean-RTPc > 312ms was 90/94 for diagnosis (vs gene-negatives and controls), 100/70 for CA vs all other LQTS and 93/88 for all symptomatic vs asymptomatic LQTS.
Conclusions: Mean Holter RTPc measured with this method is superior to the 12-lead ECG in diagnosis of LQTS and also identifies those at risk of symptoms and cardiac arrest.

O1807 - ANXIETY AND DEPRESSION IN CHILDREN WITH PACEMAKERS (PM) AND IMPLANTABLE CARDIOVERTER DEFIBRILLATORS (ICD)

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Background: The implantation of a PM or an ICD during childhood may reduce quality of life and lead to depression and anxiety disorders. The aim of this study was to evaluate the incidence of depression and anxiety symptoms in patients with implanted cardiac rhythm devices.
Methods: We analysed data from 86 pts. (63 pts. with PM, 23 with ICD) aged 6-19 (mean 12.88 ± 3.96 years), 42 female and 44 male pts. Depression was assessed by using the 20-item Center for Epidemiological Studies Depression Scale for Children (CES-DC). Anxiety disorders were analysed with the 41-item Screen for child anxiety related emotional disorders (SCARED). Both instruments were used as self- and proxy-report. The results were compared to age-matched control groups.
Results: Adolescent patients with implanted cardiac rhythm devices show higher prevalence of anxiety compared to the healthy controls (26.8% vs. 9.9%). Especially adolescent girls with PM or ICD have a higher risk to develop an anxiety disorder in comparison to the normal population (35.3% vs. 11.3%). The self- and the proxy reports did not show a difference in the incidence of depressive symptoms between children with a PM or ICD compared to the control groups. There was no statistical significant difference in the prevalence of anxiety and depression between children with implanted PM or ICD respectively.
Conclusions: Children with implanted cardiac rhythm devices have an increased risk to develop psychological diseases especially anxiety disorders. However also a quarter of the patients showed evidence of depression. Therefore in line with the regular check-ups patients and their parents should be advised and supported in terms of psychological disorders. Screening tests on the basis of standardised questionnaires should be an important part of complex care of these young patients. If necessary further diagnostics and psychotherapy should be taken into consideration.

O1978 - COGNITIVE REQUIREMENTS FOR AUTOMATIC EXTERNAL DEFIBRILLATOR USE. ARE WE STARTING TOO YOUNG

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The purpose is to assess pediatric patients' cognitive ability to appropriately perform CPR and utilize an AED on a human during cardiac arrest.
Methods: A search for data regarding pediatric use (<18 years) of AED's and cognitive ability to perform similar emergent tasks was performed. Data were assimilated and discussed by pediatric electrophysiologist and psychologist. Comparisons were made between emergent duties (exit row seating on plane) and AED use with regard to cognitive and societal beliefs as to when children are prepared to perform these tasks.
Results: The psychological impact of performing CPR and appropriate use of AED requires high levels of cognitive processing. The three cognitive decisions that a child must make to perform these tasks on a human being include: recognizing cardiac arrest, training in CPR and AED and ability to appropriately performing CPR and utilize the AED. Studies of executive function of the developing brain reveal that younger cohorts (7-12) have single factor modeling (1 task) while older cohorts (13-15 years) have fully developed three factor model (3 tasks) consistent with higher level of executive function development.
Conclusion: The cognitive requirements necessary to appropriately perform CPR and utilize an AED on a human being includes the executive function of the brain to perform well in a three factor model, (recognize cardiac arrest, be trained in and appropriately use both CPR and AED). The group recommends that prior to teaching this necessary skill that one must be cognizant of the child's cognitive developmental status and should wait until the child is at least 13 years of age.

O2053- CLINICAL AND HEMODYNAMIC ASSOCIATIONS IN INFANTS, TODDLERS AND PRESCHOOL CHILDREN

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Aim: To disclose the peculiarities of formation of tachycardia-induced cardiomyopathy syndrome in children of different ages with the help of clinical and hemodynamic data.
Materials and Methods: A total of 158 children with idiopathic arrhythmias aged 0 to 7 years were examined. Nosological groups of arrhythmias included WPW syndrome (n = 82), atrial ectopic tachycardias (n = 44), atrial flutter (n = 11), AV reentry junctional tachycardias (n = 7), and ventricular tachycardias (n = 13). Age groups comprised infants (n = 73), 1–3-year-old children (n = 43), and 3–7-year-old children (n = 98).
Results: The predominant types of arrhythmias were paroxysmal tachycardias in 3–7-year-old children ($\chi = 11.077$, $p = 0.001$) and incessant tachycardias in infants ($\chi = 13.030$, $p < 0.001$). The frequency of paroxysmal tachycardia episodes was higher in infants compared with 1–3-year-old ($p = 0.028$) and 3–7-year-old children ($p < 0.001$). Based on the echocardiography findings, signs of tachycardia-induced cardiomyopathy found more often in older children compared with the younger patients ($\chi^2 = 12.312$, $p = 0.002$). On the contrary, the presence of heart failure was more typical for infants than for 1- to 7-year-old patients ($F = 44.117$; $p = < 0.001$).
Conclusions: Arrhythmogenic heart remodeling occurred more frequently in children aged 3 to 7 years. Clinical signs of heart failure in infants with arrhythmias preceded structural changes and functional abnormalities in the intracardiac hemodynamics indices. The preservation of normal sizes of cardiac chambers in the presence of high heart rates probably resulted in the diastolic dysfunction in infants. The factors leading to these hemodynamic changes were the high average daily heart rate, tendency of tachycardia to permanent and continuously recurring course, and high frequency of paroxysmal tachycardia episodes in infants.

O2120 - THE CLINICAL COURSE AND PROGNOSIS OF MULTIFOCAL ATRIAL TACHYCARDIA IN CHILDREN

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Background: Multifocal atrial tachycardia (MAT) is known to have favorable outcome. However pediatric clinical data is insufficient. We report 33 patients with MAT at our institution to ascertain clinical course of MAT and verify possible prognostic factor.
Materials and methods: Medical records of 33 patients diagnosed with MAT from 1997 to 2015 were reviewed. Demographic information, underlying diseases, echocardiographic data were evaluated. Unfavorable outcome defined as mortality and arrhythmia duration greater than 1year and arrhythmia control rate were assessed as outcome.
Results: In 33 patients (19 boys and 14 girls), median age at diagnosis was 1.7 months (range, 0 day to 14.2 years). Twenty seven patients (81.8%) were diagnosed in infancy or neonatal period. Fourteen patients (42.4%) had structural heart disease, including five with hypertrophic cardiomyopathy. Eight had lung disease and six had syndrome all belongs to RASopathy. Two patients developed polymorphic ventricular tachycardia several years later with RyR2 gene mutation confirmed. Median kinds of anti-arrhythmics administered per a patient were 2.4. Direct-current cardioversion or adenosine was tried in eight and nine patients, but was never effective. MAT in 26 patients (83.9%) had been controlled within average 1.2 years. Four mortality cases were reported (3 cardiopulmonary and 1 non-cardiopulmonary mortality). Control rate was higher in infantile group (84.6%) than non-infantile group (66.7%), but was not statistically significant. Accompanying comorbidities was associated with lower control rate without statistical significance. However idiopathic infantile patients had significantly lower rate of adverse outcome than the others ($p = 0.03$).
Conclusions: MAT mostly affects infants and usually has favorable prognosis especially in idiopathic infantile cases. However patients with underlying comorbidities or older than one-year of age at diagnosis have varying clinical course. Long-term follow-up for further arrhythmia and thorough survey for underlying conditions are needed in these patients.

O2386 - CLINICAL OUTCOMES OF LEFT CARDIAC SYMPATHETIC DENERVATION A MULTI CENTRIC STUDY

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Background: Left cardiac sympathetic denervation (LCSD) has been used for many years in treating refractory ventricular arrhythmias. We report a multicentre experience in performing LCSD.
Materials and methods: 29 consecutive patients were included, who underwent LCSD since Nov-2011 until Nov-2016.

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Syncope, aborted cardiac arrest (ACA) and appropriate ICD discharge, were classified as cardiac events (CE).
Results: Median age was 11,8 years (range 1.0-52.1), weight was 30.0 kg (range 7.8-100), 16(55,2%) were male. Diagnoses included 15(51,7%) patients with LQTS (9 (31,1%) with JLN), 9(31,1%) with CPVT, 1(3,4%) with histiocytoid cardiomyopathy, 1(3,4%) with Emery-Deifuss, and 3(10,3%) with IVF. Prior to LCSD, 14(48,3%) had experienced ≥10 CE, 9(31%) had 5-to-9 CE, and 5(17,2%) had 1-to-4 CE, and 1(3,4%) had no-CE, with an overall median of 11,5(0-87) CE. All patient had tried maximal dose of beta-blockers (BB) before LCSD, 6 were on BB-flecainide, 1 on BB-mexilitine and 1 had tried BB, amiodarone, sotalol and lignocaine. 20(69%) had an ICD before LCSD, all for secondary prevention. Video assisted thorachoscopic approach (VATS) could be performed in 28(96,6%) patients. Only the smallest patient (7,8 kg) underwent a postero-lateral thoracotomy approach as the subpleural ICD coil interfered with VATS ports incisions. Intraoperative complications included 1 VF episode. Long-term side effects included 2 patients with a transient Horner's syndrome (HS) and 1 with permanent HS. Over a median follow up of 42 months (range 1,7-57,5), 21(72,4%) patients had no breakthrough of CE, 5(17,2%) had 1-4 BCE, 2(6,9%) had 5-to-9 BCE and 1(3,4%) had >10 BCE. Overall, there was a significant reduction of CE after LCSD (11.5 (0-87) to 0(0-7); p:0.001) Figure1.
Conclusions: LCSD is a safe and effective technique with few complications in drug refractory ventricular arrhythmias due to channelopathies and other cardiomyopathies. VATS approach is a safe and feasible procedure but may be difficult in very small patients. A significant reduction of BCE has been seen in the majority of patients.

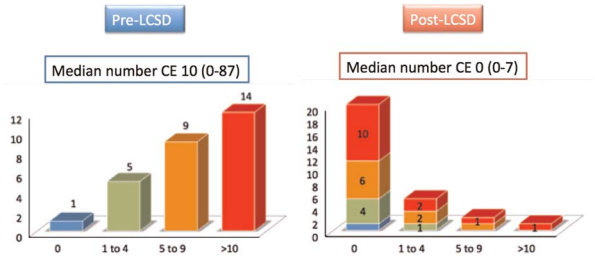

Figure 1.
21(72,4%) patients had no breakthrough of CE, 5 (17,2%) had 1-4bce, 2 (6,9%) had 5-to-9 and 1(3,4%) had >10 BCE. There is a significant reduction of EC after LCSD: Median (11.5(0-87) to 0 (0-7); p:0.001).

O2406 - RISK STRATIFICATION FOR ARRHYTHMIAS IN PEDIATRIC HYPERTROPHIC CARDIOMYOPATHY

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Background/Hypothesis: Risk stratification for sudden arrhythmic death in pediatric hypertrophic cardiomyopathy (HCM) remains challenging and guidelines for adult HCM are not validated in children.
Objective: To evaluate clinical predictors for arrhythmias in patients with pediatric onset HCM.
Materials and Methods: 40 patients with a diagnosis of HCM between 0 and 18 years of age underwent a detailed clinical evaluation including genetic testing, electrocardiogram, 24-hour Holter recording, cardiopulmonary exercise testing, transthoracic echocardiography (TTE), and magnetic resonance imaging (MRI) with contrast at a follow-up time of 11 ± 6 years. Arrhythmic events were defined as non-sustained or sustained supraventricular or ventricular tachycardia or appropriate implantable cardioverter defibrillator discharge. Clinical characteristics of patients with and without arrhythmic events were compared.
Results: Data are depicted in the Table. Genetic diagnosis was Noonan syndrome or mutations in the sarcomere. Patients with arrhythmias had disease onset earlier in age, significant thicker left ventricular wall thickness on TTE, higher extracellular volume fraction on MRI T1-map as an estimate for interstitial fibrosis, and were more likely to carry a mutation in the troponin T (TNNT) gene. There was no significant difference between groups with regards to genetic diagnosis, the presence of late gadolinium on MRI as an estimate for focal fibrosis, history of syncope, family history of sudden cardiac death, left ventricular outflow tract obstruction and enlarged left atrium.
Conclusion: Risk factors for arrhythmias included early disease onset, magnitude of myocardial hypertrophy and interstitial fibrosis, and the presence of TNNT mutations. This underlines the importance of a thorough clinical evaluation including MRI imaging with contrast and genetic testing in patients with pediatric onset HCM.

Table.

	No arrhythmias	Arrhythmias	p-value
Patients (N)	34	6	NS
Noonan HCM/sarcomere HCM)	12/22	1/5	
Gender (male/female)	19/15	4/2	NS
Sarcomere HCM pts	13/9	3/2	
Age at diagnosis (years)	5.6 ± 6.5	1.8 ± 2.9	0.034
Sarcomere HCM pts	7.6 ± 6.7	2.2 ± 3	0.016
LVWT (z-score)	5.0 ± 1.8	7.0 ± 1.0	0.009
Sarcomere HCM pts	5.3 ± 2.0	6.8 ± 0.8	0.015
ECV (%)	29 ± 4	35 ± 2	0.037
Sarcomere HCM pts	29 ± 5	35	NS
LGE positive	15/27 (56%)	1/3 (33%)	NS
Sarcomere HCM pts	8/18 (44%)	1/2 (50%)	NS
TNNT positive	NA	NA	NA
Sarcomere HCM pts	1/22 (5%)	2/5 (40%)	0.08
LVOTO	20/34 (59%)	3/6 (50%)	NS
Sarcomere HCM pts	11/22 (50%)	2/5 (40%)	NS
Syncope	4/34 (12%)	2/6 (33%)	NS
Sarcomere HCM pts	4/22 (18%)	2/5 (40%)	NS
Family history SCD	2/34 (6%)	2/6 (33%)	NS
Sarcomere HCM pts	2/22 (9%)	2/5 (40%)	NS
Enlarged left atrium	13/32 (41%)	0/5 (0%)	NS
Sarcomere HCM pts	8/21 (38%)	0/4 (0%)	NS

pts: patients; NS: not significant; LVWT: left"ventricular wall thickness; ECV: extracellular volume fraction; LGE: late gadolinium enhancement; NA: not available; LVOTO: left"ventricular outflow tract obstruction; SCD: sudden cardiac death; p-value: independent t-test or chi-square.

O2454 - FINDING A NEW RHYTHM A SPECIALISED MULTIDISCIPLINARY MODEL OF CARE TO IMPROVE PSYCHOLOGICAL ADJUSTMENT IN FAMILIES OF CHILDREN WITH RARE INHERITED ARRHYTHMIA CONDITIONS

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Background and Hypotheses: Families of children with a rare inherited arrhythmia condition (IAC) face challenges associated with exposure to traumatic events, complex genetic testing options, activity restrictions, lifelong clinical surveillance, and an increased risk of sudden death. These challenges can bear heavily on children and families, yet parents often struggle to find appropriate psychological support. This prospective cohort study assesses the prevalence and predictors of psychological morbidity in families of children with an IAC.
Materials and Methods: Prior to attending a multidisciplinary IAC clinic in Sydney, Australia, parents complete measures assessing their child's quality of life (QOL) and their own symptoms of post-traumatic stress, anxiety and depression. Family functioning, social support, health literacy and unmet needs are also assessed. Follow-up assessments are completed 1-, 6- and 12-months post-clinic.
Results: Sixty-one parents from 36 families have enrolled in the study (response rate: 62%). Diagnoses include Long QT Syndrome (62%), Catecholaminergic Polymorphic Ventricular Tachycardia (29%), and Brugada Syndrome (9%). Twenty-nine percent of children have had an out-of-hospital cardiac arrest. Prior to clinic, 1 in 5 parents reported symptoms of post-traumatic stress warranting clinical intervention. Fifty-five percent of mothers and 38% of fathers perceived their child's emotional health to be at-risk. Using linear regression with generalised estimating equations to account for familial clustering, and controlling for marital status and parent age, poorer child QOL was associated with lower perceived social support (Beta = 0.85, $p < 0.01$) and greater parental psychological stress (Beta = -7.87, $p < 0.005$). Determinants of poorer family functioning were lower perceived social support (Beta = -0.03, $p < 0.0001$), greater unmet information and support needs (Beta = 0.01, $p < 0.001$), greater parental age (Beta = 0.03, $p < 0.0001$), and being a single parent (Beta = 0.60, $p < 0.0001$).
Conclusions: Children with an IAC and their parents are at increased risk of psychological difficulties. Psychological care should form part of best practice recommendations for families of children with an IAC.

O2456 - CARDIAC RHYTHM DISTURBANCES IN HETEROTAXY SYNDROME

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Background: Heterotaxy syndrome is associated with complex cardiac malformations and cardiac conduction system abnormalities. Those with right atrial isomerism (RAI) have dual sinus nodes and dual atrioventricular nodes predisposing them to supraventricular tachycardia (SVT). Those with left atrial isomerism (LAI) lack a normal sins node and are at risk of sinus node dysfunction (SND) and atrioventricular block (AVB). We report the prevalence and risk factors associated with arrhythmias in heterotaxy syndrome.
Methods: A retrospective review of all heterotaxy syndrome patients born and treated at our institution between 2000 and 2014 was performed. Patient demographics, electrocardiograms, echocardiograms and operative records were reviewed. Arrhythmia diagnosis, timing, and prevalence were noted. Atrial situs, gender, univentricular physiology, ventricular dysfunction, presence of significant atrioventricular valve regurgitation, and surgical details such as history of multiple surgeries, TAPVC repair, atrioventricular valve surgery were assessed for potential risk factors of arrhythmia development.
Results: A total of 42 heterotaxy patients were identified; 18/42 (43%) with LAI and 24/42(57%) with RAI. There were 13 deaths [LAI 4/18(10%), RAI 9/24(21%); $p = 0.33$], one of which was due to ventricular fibrillation. The median follow-up period was 5.2 ± 5.3 years. Twenty-two patients had arrhythmias during follow-up; 13/18(72%) in LAI and 9/24(31%) in RAI ($p < 0.05$). In LAI, 10/13 had SND, and 3/10 had AVB. In RAI, 5/9 had SVT, 2/9 had atrial ectopic tachycardia, and 1/9 had junctional tachycardia. Freedom from arrhythmia at 1,3,5 years of age were 23.0%, 53.3%, 68.9% in LAI, and 17.4%, 23.3%, 31.0% in RAI respectively. No risk factors for arrhythmia were identified.
Conclusions: Arrhythmias were commonly seen in heterotaxy syndrome particularly in LAI with more than 50% having arrhythmias at 3 years of age. Clear risk factors were not identified and detailed assessment of new onset arrhythmias and close follow-up are warranted in these high-risk patients.

O2658 - ELECTRO MECHANICAL DYSFUNCTION IN TUBEROUS SCLEROSIS COMPLEX ROLE FOR ARRHYTHMOGENIC RISK PREDICTION AND TREATMENT OPTIONS

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Objective: Tuberous sclerosis complex (TSC) is an inherited neurocutaneous disorder related to cardiac rhabdomyomas. Rhabdomyomas are typically multifocal and can cause flow and electrical abnormalities. Our aim was to evaluate left ventricular (LV) mechanics by using speckle tracking echocardiography (STE) and ECG features in patients with TSC and rhabdomyomas before and after a course of Everolimus (mTOR inhibitors).
Methods: We studied 16 patients with TSC (median age 1.2 years, range 0-3 years, LVEF >50%). All underwent standard echocardiographic evaluation and STE. ECG and 24 h tape date were also obtained.
Results: Diastolic parameters were significantly abnormal in our patients before and after everolimus (E/e' average: 10.9 ± 5.7 vs. 5.6 ± 3.2, $p = 0.0016$). Global LV longitudinal strain was significantly lower before everolimus ($-16.6 \pm 3.6\%$ vs. $-23.2 \pm 3.1\%$, $p < 0.0001$). LV torsion ($9.1 \pm 4.9°$ vs. $11.9 \pm 3.3°$, $p = 0.046$) was significantly impaired before everolimus. All patients had impaired cardiac repolarization (long QT) and prolonged time to diastolic peak before everolimus course.
Conclusions: Diastolic function, LV longitudinal deformation and LV torsion in patients with TSC improved after a course of everolimus. m-TOR inhibitors have a positive effect on electromechanical cardiac function. Treatment decreases cardiac dysfunction and risk for ventricular arrhythmia. This indicates the potential added value of the assessment of mechanical dysfunction in future risk stratification of TSC patients.

O2730 - COMPARATIVE ANALYSIS BETWEEN ELECTRO AND ECHOCARDIOGRAPHIC FINDINGS OF POOR PROGNOSIS AFTER TOTAL CORRECTION FOR TETRALOGY OF FALLOT

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Background: Common complications after total correction of tetralogy of Fallot (T4F), include pulmonary insufficiency and right and/or left ventricular dysfunction. They are related to ventricular arrhythmias at early ages and to sudden death. Electrocardiographic (ECG) parameters, such as QRS enlargement above 180 ms, echocardiographic (ECHO) and nuclear magnetic resonance (MRI) parameters have been identified as predictors of these events. This study aims to describe the ECG profile of patients after total correction of T4F and to compare the prevalence of ECG findings of poor prognosis with ECHO findings of residual lesions and ventricular dysfunction, in these patients.

Material and Methods: Retrospective analysis of a database from a Paediatric Cardiology Reference Service of the state of Pernambuco. Among 169 T4F echocardiographic diagnoses, 30 met all the inclusion criteria for analysis of the electrocardiographic profile. Statistical analysis was performed, with $p < 0.05$ being considered significant.

Results: ECG and ECHO were performed on average 4.5 years after corrective surgery. Only one patient presented QRS > 180 ms and six had QRS > 160 ms. The mean QRS duration in the sample was 116 ms. Among patients with RV dysfunction, 66% had QRS > 160 ms, against 15% of those without RV dysfunction ($p = 0.03$). Among the patients with RV dysfunction on ECHO, 66% had a U wave on the ECG ($p < 0,01$).

Conclusion: A lower mean QRS duration was found in the studied sample in comparison to other large series of the literature. This may reflect the repair technique, which did not use a transannular patch. A statistically significant relationship was found between the enlarged QRS interval and ventricular dysfunction. The presence of a U wave was related to right ventricular dysfunction. ECG and ECHO can provide important parameters for the follow-up of patients after total correction of T4F, especially in centers with less technological resources.

O2740 - OUTCOME OF TRANSVENOUS LEAD EXTRACTION IN CHILDREN A SIXTEEN YEAR PAEDIATRIC CASE SERIES.

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Background: Lead extraction is increasingly required to revise chronic transvenous (TV) pacemakers (PM) and defibrillators (ICD). Children pose a particular challenge, as they are implanted at a young age, will require system revisions and have increased related lead failures. Leaving faulty leads in situ avoids all risk but increases the risk of venous obstruction. We report our multicenter experience over 16 years.

Materials and Methods: Retrospective study of outcomes in children ≤16 years of age requiring TV lead extractions between Jan-2000 and Dec-2016.

Results: There were 30 procedures in 28 patients. 14 males(50%). Mean age at time-of-device-implant: 7.5 years(IQR 4.8-11.1), mean age at time-of-lead-extraction: 11.6 years(IQR 9.8-13.9) and a total of 39 leads were extracted. The indication for device implant included CCHB 15, LQTS 6, CPVT 3, BrS 1, TAPVD 1, sick sinus syndrome (SSS) and repaired muscular VSD 1 and JET 1. A total of 20 pacemakers included 14 VVI, 4 DDD, 1 CRT-P, and 1 AAI and 9 ICDs, comprising 6 single and 3 dual chamber. The types of leads and age-at-time-of-extraction comprised 9 atrial leads (mean lead-age 25.3 months; IQR 6.5-36.8), 21 pace/sense leads (mean lead-age 63.8 months; IQR 21-105) and 9 defibrillator leads (mean lead-age 20.1 months; IQR 10.3-33.9). The reason for extraction was lead failure in 18 patients (61%), upgrades/system in 6 cases(21%) and 6 extractions(18%) were due to infection. Complete procedural success was achieved in 27 cases(89%), partial success in 1(4%) and failure in 2 cases(7%). Advanced Laser or mechanical rotor was required in 15(52%) cases and combined locking stylets with manual traction in 14(48%). No major complication or 30 day mortality occurred.

Discussion: Lead extraction can be successfully achieved in 90% of children without complications or 30 day mortality. However, it should be reserved for specialist congenital cardiac centers with careful case selection in the very young.

HUMANITARIAN

O1227 - THE HUMANITARIAN FOOTPRINT CONTRIBUTIONS OF GLOBAL PEDIATRIC CARDIAC SURGERY TO THE HUMAN DEVELOPMENT INDEX OF LOW AND MIDDLE INCOME COUNTRIES

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Background: International health organizations endorse global campaigns for humanitarian interventions such as vaccinations and water sanitation due to their proven cost-effectiveness, but remain reluctant to support seemingly expensive global interventions in surgery. We measured the footprint of humanitarian pediatric cardiac surgery in low-middle income countries (LMIC) by gains in the main components of the Human Development Index (HDI): Life Expectancy (LE), Median Years of Schooling (MYS) and Gross National Income per capita (GNIpc). We hypothesized that humanitarian pediatric cardiac surgery would lead to gains in HDI and be categorized as a cost effective intervention.

Material and Methods: After assessing total administrative, logistical and clinical costs incurred by our organization during 2015 and obtaining current HDI indicators for each country visited, independent reviewers calculated the potential incremental difference in these indicators after our intervention. A cost per surgery in USD$ was ascribed to these differences in HDI and a cost effectiveness ratio in disability adjusted life years (DALY) averted was determined for each patient.

Results: During 2015, 444 patients underwent 470 operations in 10 LMIC (mortality 7.8%) at a cost per patient operated of $7,362. After excluding deceased (n = 35), adult (n = 13) and non-cardiac (n = 6) patients, incremental differences in HDI indicators were estimated for the remaining 390 patients. Our global interventions resulted in 17,258.97 DALY averted of LE; 2,308 extra years of schooling and USD$225,639,139 at purchasing power parity of GNIpc gained, representing an average of 44.25 DALY averted of

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LE; 5.9 extra years of schooling and $578,561 of GNIpc gained per patient at a cost of $166 per DALY averted.
Conclusions: Humanitarian pediatric cardiac surgery in LMIC improves human development indices and is as cost-effective as other subsidized humanitarian interventions. Global health organizations should encourage and support sustainable approaches to humanitarian pediatric cardiac surgery.

O1500 - GLOBAL BURDEN OF CONGENITAL HEART DISEASE

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Background: There is a large knowledge gap in understanding the global burden of congenital heart disease (CHD). The Institute for Health Metrics and Evaluation (IHME) has collected data worldwide and estimated mortality and morbidity of CHD from 1990 to 2015.
Methods: All available data sources on fatal and non-fatal CHD from 195 countries and territories were systematically reviewed for inclusion in epidemiologic models. Cause of Death Ensemble modeling and DisMod-MR 2.0 were used to produce consistent estimates of mortality and prevalence, respectively, including corresponding uncertainty. Using the publically-available Global Burden of Disease (GBD)-Compare visualization tool (http://vizhub.healthdata.org/gbd-compare), we evaluated CHD burden by seven GBD super-regions. Mortality estimates are available for aggregate CHD; non-fatal estimates are further divided into: critical, severe, and less severe CHD.
Results: Annual mortality (Table-1) from CHD worldwide is estimated to be 303,282 (95% CI 268,772-335,386); 24% of deaths occur in sub-Saharan Africa. Mortality rate in sub-Saharan and North Africa is 7-8 per 100,000 compared to 1 per 100,000 in high-income countries. The mortality rate due to CHD in infants decreased significantly from 1990 to 2015 only in high and middle-income countries. The global prevalence of CHD (Table-2) is 48.9 million (95% CI 34.3-72.8 million). Prevalence, expressed as a percent, is lower in low-income countries (0.2-0.6%) compared to high-income countries (0.9%), suggesting a greater percent of deaths from other causes. The global prevalence of critical, severe and less severe CHD for all ages is 0.002% (0.001%-0.003%), 0.05% (0.04%-0.08%), and 0.59% (0.39%-0.91%), respectively. The prevalence for critical and severe CHD drops below 0.01% and 0.1%, respectively, in all regions except high-income countries by one year of age. Uncertainty estimates are highest in low-income countries.
Conclusions: The novel IHME GBD Compare Tool facilitates visualization of best estimates of GBD data, which improves our understanding of global CHD burden. It provides critical information to help address global inequalities.

Table 1. Congenital heart Disease mortality

Location	< 1 year		All Ages	
	Percent of Deaths	Rate of Death (per 100,000)	Percent of Deaths	Rate of Death (per 100,000)
1990				
Global	3.16% (2.27-4.47)	4047 (2962-5009)	0.81% (0.60-1.13)	7.32 (5.37-10.29)
High-income	13.09% (9,85-14.15)	2933 (2080-3293)	0.31% (0.25-0.33)	2.69 (2.17-2.85)
Central/Eastern Europ and Central Asia	8.31% (6.59-9.24)	5832 (4403-6562)	0.46% (0.38-0.50)	4.89 (4.0-5.25)
Latin America and Caribbean	3.73% (3.26-4.95)	3155 (2831-3821)	0.99% (0.89-1.26)	5.81 (5.24-7.40)
North Africa and Middle East	5.79% (3.51-10.81)	6737 (4334-10557)	2.06% (1.29-3.78)	14.81 (9.28-27.63)
South Asia	1.73% (0.99-2.47)	3865 (2119-5562)	0.72% (0.45-0.95)	7.64 (4.82-10.12)
Southeast Asia, East Asia, and Oceania	5.25% (3.71-7.89)	4540 (3316-5651)	1.11% (0.81-1.65)	8.31 (6.06-12.36)
Sub-Saharan Africa	1.55% (0.84-2.77)	2566 (1445-4316)	0.71% (0.41-1.33)	9.90 (5.67-18.70)
2015				
Global	4.39% (3.69-5.11)	2764 (2223-3248)	0.54% (0.48-0.60)	4.11 (3.65-4.55)
High-income	10.37% (9.33-12.10)	788 (694-953)	0.12% (0.11-0.13)	1.05 (0.98-1.20)
Central/Eastern Europ and Central Asia	9.55% (8.06-11.26)	2813 (2337-3326)	0.19% (0.17-0.22)	2.25 (2.0-2.56)
Latin America and Caribbean	9.26% (7.79-10.22)	2791 (2313-3149)	0.61% (0.51-0.66)	3.56 (2.99-3.88)
North Africa and Middle East	10.63% (8.16-14.03)	5257 (4006-6832)	1.48% (1.22-1.90)	7.82 (6.40-9.92)
South Asia	2.75% (1.57-3.99)	3038 (1564-4783)	0.59% (0.41-0.75)	4.47 (3.15-5.65)
Southeast Asia, East Asia, and Oceania	11.12% (8.99-13.22)	2778 (2120-3402)	0.49% (0.40-0.57)	3.27 (2.71-5.65)
Sub-Saharan Africa	2.57% (1.67-3.70)	2182 (1445-3231)	0.92% (0.64-1.26)	7.69 (5.36-10.46)

Table 2. Prevalence of congenital heart disease: 2015.

Location	All Severities		Less Severe	
	< 1 year	All Ages	< 1 year	All Ages
Global	0.78% (0.51-1.21)	0.65% (0.43-1.00)	0.68% (0.45-1.06)	0.59% (0.39-0.91)
High-income	1.02% (0.69-1.55)	0.91% (0.61-1.39)	0.79% (0.54-1.22)	0.75% (0.50-1.26)
Central/Eastern Europe and Central Asia	1.05% (0.73-1.57)	0.86% (0.60-1.28)	0.92% (0.64-1.37)	0.79% (0.55-1.18)
Latin America and Caribbean	0.71% (0.44-1.13)	0.57% (0.35-0.92)	0.61% (0.38-0.99)	0.53% (0.33-0.85)
North Africa and Middle East	0.77% (0.52-1.21)	0.60% (0.40-0.93)	0.69% (0.47-1.08)	0.57% (0.39-0.88)
South Asia	0.63% (0.37-1.06)	0.37% (0.21-0.63)	0.58% (0.34-0.99)	0.36% (0.21-0.62)
Southeast Asia, East Asia, and Oceania	1.08% (0.73-1.64)	0.93% (0.64-1.42)	0.93% (0.64-1.43)	0.85% (0.59-1.30)
Sub-Saharan Africa	0.53% (0.35-0.84)	0.22% (0.14-0.36)	0.48% (0.31-0.76)	0.22% (0.13-0.35)

Location	Severe		Critical	
	< 1 year	All Ages	< 1 year	All Ages
Global	0.08% (0.06-0.13)	0.05% (0.04-0.08)	0.01% (0.01-0.02)	0.002% (0.001-0.003)
High-income	0.20% (0.13-0.29)	0.15% (0.10-0.22)	0.03% (0.02-0.05)	0.009% (0.005-0.014)
Central/Eastern Europe and Central Asia	0.11% (0.08-0.17)	0.06% (0.04-0.09)	0.02% (0.01-0.03)	0.002% (0.001-0.004)
Latin America and Caribbean	0.08% (0.06-0.13)	0.04% (0.03-0.07)	0.01% (0.01-0.02)	0.001% (0.000-0.001)
North Africa and Middle East	0.07% (0.05-0.11)	0.03% (0.02-0.05)	0.01% (0.01-0.02)	0.001% (0.000-0.001)
South Asia	0.04% (0.02-0.06)	0.003% (0.002-0.001)	0.01% (0.00-0.01)	0.000% (0.000-0.000)
Southeast Asia, East Asia, and Oceania	0.13% (0.09-0.19)	0.08% (0.05-0.12)	0.01% (0.01-0.02)	0.002% (0.001-0.003)
Sub-Saharan Africa	0.04% (0.03-0.07)	0.002% (0.001-0004)	0.01% (0.00-0.01)	0.000% (0.000-0.00)

O1739 - HUMANITARIAN MODEL TO RESCUE THE INVISIBLE CHILD FREE CARDIAC CARE FOR CONGENITAL HEART DISEASE IN INDIA

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Background: Overall, global childhood mortality decreased by 50% but death and disability from congenital heart disease (CHD) increased progressively in low-/middle-income countries (LMICs) over the past 2 decades. Of the 1·35 million children born each year with CHD, 90% do not have access to diagnostics or adequate care. Communicable diseases dominate the healthcare budget of LMICs, whereas most children with CHD remain undiagnosed and untreated. We present a unique model of preventive, palliative, and curative cardiac care wherein screening, diagnostic, medical & surgical services are provided free of charge.
Materials and Methods: Programmatic and clinical outcomes at Sri Sathya Sai Sanjeevani Hospital (SSSSH) in Naya Raipur were analyzed for 2015 & 2016. All medical, surgical, consultative and ancillary services are rendered totally free of cost to all those who arrive at SSSH.
Results: A total of 8530 children with CHD were evaluated in the out patient department from Jan 2015 to Dec 2016, out of which 2188 were operated. The 30- day mortality for year 2015 was 2.5%, which dropped to 1.3% in year 2016. The STS-EACTS distribution and corresponding mortality for the years 2015 and 2016 are presented in the table (Table 1). The average cost per

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surgery was INR 67,000 (USD 1000). Many innovative measures are used to keep the cost of treatment low, one of which is training parents to assist the medical team in taking care of their children in the postoperative period.
Conclusions: Three hundred thousand children with congenital heart disease are born each year in India. A tiny fraction of these are treated at present. SSSSH is making a small but significant contribution by addressing the national burden of congenital heart disease.

Table 1.

	2015		2016	
STS-EACTS CAT	**NUMBER (%)**	**MORTALITY (%)**	**NUMBER (%)**	**MORTALITY (%)**
CATEGORY 1	562 (56.2%)	3 (0.5%)	679 (57%)	5 (0.7%)
CATEGORY 2	374 (37.4)	9 (2.4%)	393 (33%)	2 (0.5%)
CATEGORY 3	018 (1.8%)	7 (38.8%)	026 (2.2%)	-
CATEGORY 4	046 (4.6%)	6 (13.4%)	088 (7.4%)	8 (9.1%)
CATEGORY 5	-	-	002 (0.1%)	1 (50%)

O2273 - SUCCESSFUL MODEL OF A SUSTAINABLE PROGRAM FOR LOW INCOME PEDIATRIC CHILDREN WITH CONGENITAL HEART DISEASES IN DEVELOPING COUNTRIES

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Background: Social Program Give a Life is sustainable program in pediatric cardiac care, for the most needed children in Colombia. Eleven cities are visited every year, looking for children without health insurance who need cardiac care. 2148 children were treated in 13 years of work. 3500 children per year receive free evaluation. The program is supported by local donations and the institution philosophy. Free surgical treatment, transportation and housing is provided to all children and part of their relatives, and medical follow up after the discharge is provided by local allies. The aim is to describe the outcomes of these children undergone to surgery to repair of congenital heart disease compare with all ensured ones matched by RACHS-1 categories 1-3.
Methods: Cohort of patients who belong to the Social Program, from January 2010 to October 2016 who were treated by surgery. Standard statistical methods were used to describe the patient´s characteristics, operative variables, and post operative outcomes.
Results: In this study period 3046 patients were operated by the surgical group, the 15.7% (478/3046) belongs to social program. The 89% (425/478) were between 1 to 18 years old. Procedures in RACHS-1 risk 2, 39.7%, follow by risk 1, 26.4%, and risk 3, 24.7%. Median ICU stay 2 days, median in hospital stay 2 days. Global mortality 1%. Survival at 30 days 99%. In table one are describe the results compare with the rest of the cohort.
Conclusions: Referral centers can develop sustainable program and achieve the same results and guarantee medical follow up for noninsurance patients in developing countries as part of their mission, with no differences in the clinical outcomes. This Initiatives could be reproduced in other countries or centers to help the most needed.

Table 1. Differences between Social Patients and Insurance Patients.

Variable	Social Program n = 434	Not Social Program N = 2022	P Value
Male %	49.1	52.2	0.001
Premature %	12,9	19.7	0.004
Malnoursih %	29.7	31.5	0.45
Chromosome Anomalies %	3.4	15.7	0.001
Medical Diseases %	11.3	17.8	0.001
Reoperations %	5.3	5.4	0,93
Renal Failure %	2.1	3.4	0,15
Neurological %	2,8	2.7	0,91
Pulmonary %	13.1	17.7	0,05
Cardiac %	16,1	18.6	0,22
Mortality %	1%	1%	0.89

IMAGING

O1052 - QUANTIFICATION OF BLOOD FLOW BY AUTOMATED THREE DIMENSIONAL REAL TIME VOLUME COLOUR FLOW DOPPLER ECHOCARDIOGRAPHY IN PEDIATRIC ATRIAL SEPTAL DEFECTS

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Objective: Quantification of mitral and aortic valve flows and left ventricular volumes has been validated in adult hearts using real-time three-dimensional (3-D RT-VCFD) echocardiography. In this study, we sought to validate the accuracy of this novel technology in smaller pediatric hearts, across mitral (MV), aortic (AV), pulmonary (PV) and tricuspid (TV) valves and for right (RV) and left (LV) ventricles during percutaneous atrial septal defect (ASD) device closure.
Methods: 45 studies were prospectively performed in 25 children while under general anesthesia and breath-holding technique. 3-D colour flow Doppler across each valve and full volume of both ventricles with ECG-gating were acquired using the Acuson SC2000 (Siemens, CA) machine for 3 beats before and after ASD closure. The 3-beat averaged flows and volumes were measured using a dedicated offline 3-D Flow computational program. Qp/Qs was calculated by the Fick method.
Results: The mean body weight was 30.3 ± 15.4 kg and height 129.6 ± 22.0 cm. The mean Qp/Qs was 1.7 ± 0.5 before closure. Valve flow correlation was high between the PV and TV ($R = 0.96$, $p < 0.001$) and between the AV and MV ($R = 0.91$, $p < 0.001$). There was good correlation between the RV stroke volume and flow across the TV ($R = 0.95$, $p < 0.001$) and PV ($R = 0.88$, $p < 0.001$). The correlation was moderate between the LV stroke volume and the AV ($R = 0.70$, $p < 0.001$) and MV ($R = 0.81$, $p < 0.001$). Qp/Qs correlated with PV:AV flows ($R = 0.82$, $p < 0.001$) and TV:MV flows ($R = 0.84$, $p < 0.001$). There was moderate correlation between Qp/Qs and RV:LV volumes ($R = 0.62$, $p < 0.01$).
Conclusion: Flow measurement using 3-D RT-VCFD echocardiography is feasible and reliable in children in the left and right

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sides of the heart. The highest correlation was between valves and ventricular volumes within the same side of the heart. This technique provides a non-invasive alternative to historically invasively acquired hemodynamic data.

O1217 - MONITORING THE DEVELOPMENT OF RIGHT VENTRICULAR HEART FAILURE USING HYPERPOLARISED 13C MAGNETIC RESONANCE IMAGING

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Background: Right heart failure is common in patients with congenital heart disease, but the evaluation of right ventricular function is complicated and inaccurate and the correct timing of intervention is a matter of continuous dispute. Current diagnostics focus on alterations in anatomical and physiological parameters. Magnetic resonance hyperpolarisation, using pyruvate as an active metabolic tracer, is a novel technique that can image the metabolism in tissues non-invasively in real-time. We hypothesised that the metabolic alterations known to occur in heart failure can be assessed and monitored with this technique making it a potential powerful tool in heart failure diagnostics.
Materials and Methods: At baseline five female 5 kg piglets were subjected to banding of the pulmonary trunk. Thereby, progressive heart failure was induced by natural growth. At baseline and every four weeks for 16 weeks the animals were assessed with hyperpolarised pyruvate, conventional MR imaging, 4D echocardiography and blood sampling. Finally, at week 16 cardiac physiology was invasively assessed with conductance catheter technique. End-point data was compared to a weight matched control group.
Results: At 16 weeks right ventricular end systolic pressure-volume relationship, dP/dt_max and preload recruitable stroke work were increased ($p < 0.05$), while the left ventricle exhibited significant diastolic impairment. The myocardial lactate/bicarbonate ratio, depicting the balance between anaerobic and aerobic metabolism, increased from 0.036 to 0.10 ($p = 0.004$). No changes were found in the functional echo-derived right ventricular parameters such as TAPSE, FAC and strain.
Conclusions: At 16 weeks, the animals had developed compensated right heart failure accompanied by a significant impairment of left ventricular diastolic function. Heart failure at this level was not detectable by neither conventional MR imaging nor echocardiography whereas hyperpolarisation data showed an increase in lactate/bicarbonate ratio indicating a shift towards anaerobic metabolism. In this study hyperpolarisation was capable of detecting developing right heart failure prior to conventional imaging modalities.

O1230 - DEVELOPMENT OF SYSTOLIC VENTRICULAR FUNCTION AFTER THE FONTAN OPERATION

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Background/Hypothesis: The aim of our study was to investigate the development of systolic ventricular function in a single center Fontan cohort from preoperatively until five to ten years after the Fontan operation. Our secondary aim was to investigate the functional impact of AV valve and semilunar valve regurgitation, ventricular morphology and mechanical dyssynchrony.
Materials and Methods: We analyzed digitally stored echocardiography studies from three time points in 49 eligible patients: shortly before Fontan completion, postoperatively and five to ten years after. Systolic function was measured preoperatively and late post-Fontan by visual assessment, by fractional area change (FAC), by global longitudinal strain (GLS), and by non-septal longitudinal strain (LSans).
Results: Visually assessed ventricular function was declining by 0.08 ± 0.08 grades per year (Chi square $p < 0.001$). In contrast, FAC, GLS and LSans were unchanged from pre-Fontan until late post-Fontan for the entire group and independently of left or right ventricular morphology. Pre-operative valve incompetency did not change preoperative FAC, GLS or LSans. Linear regression analysis did not show significant influence of either ventricular morphology, gender, age at Fontan operation or post-operative valve incompetency on yearly change of FAC, GLS or LSans. Mechanical dyssynchrony was present in an equally small subgroup of patients before and late after Fontan completion.
Conclusion: Systolic ventricular function in the single ventricle circulation show remarkable stability through the first five to ten years after the Fontan operation. Neither ventricular morphology nor valve incompetency did induce significant functional deterioration. We did not find indications of substantial mechanical dyssynchrony before or late after Fontan completion.

O1331 - INCREASED WAVE REFLECTION INDICES IN YOUNG ADULTS FOLLOWING END TO END COARCTATION REPAIR COMPARED WITH SUBCLAVIAN FLAP REPAIR

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Introduction: The risk of hypertension in adulthood following aortic coarctation (CoA) repair remains high, with controversy on whether resection/end-end anastomosis (E-E) or subclavian flap repair (SFA) constitutes the best surgical approach. Routine echo measures do not identify subjects at risk of hypertension.
Hypothesis: We hypothesised that the novel technique of Wave Intensity Analysis (WIA) would reveal increased wave reflection indices in the E-E group.
Methods: Routine echo assessment of LV function and arch anatomy was performed in 37 patients with repaired CoA (20 SFA, 17 E-E). In addition, WIA was undertaken at the level of the proximal transverse arch using Doppler velocity measures and the aortic distention waveform calibrated to brachial blood pressure, with determination of the intensities of main forward and backward (i.e. reflected) compression waves (FCW and BCW), their pressure effects and the local reflection coefficient (BCW:FCW pressure change ratio).
Results: Patient demographics were similar between groups, but SFA patients had a greater age at follow-up (27 ± 5 vs 19 ± 6yrs, $p < 0.001$), a higher incidence of transverse arch hypoplasia at time of surgery (20 ± 40 vs 0%, $p < 0.04$), and a higher distal transverse arch z-score at follow-up (0.7 ± 1.3 vs -0.1 ± 0.9, $p = 0.04$).

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No differences were evident between SFA and E-E groups in LV dimensions, calculated LV mass, LV systolic function or systolic blood pressure (133 ± 13 vs 130 ± 11 mmHg, p = 0.5). However, while FCW intensities were not different between groups, E-E patients had a higher BCW intensity (p = 0.02), a greater BCW-related pressure increase (16 ± 6 vs 11 ± 5 mmHg, p = 0.02) and higher reflection coefficient (0.41 ± 0.14 vs 0.28 ± 0.11, p = 0.02).
Conclusions: Young adults following E-E repair of CoA have elevated wave reflection indices in the proximal transverse arch, and smaller distal arch z-scores compared with those repaired with SFA. These finding have important implications for development of hypertension in later life.

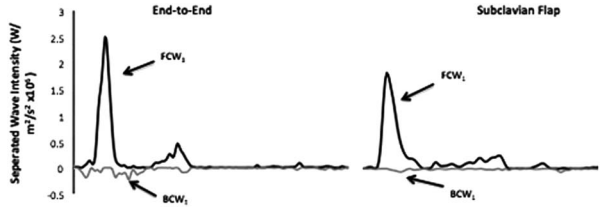

Figure.

O1386 - EVOLUTIVE ECHOCARDIOGRAPHIC ASSESSMENT OF THE RIGHT VENTRICULAR FUNCTION IN PAEDIATRIC PATIENTS WITH REPAIRED TETRALOGY OF FALLOT MYOCARDIAL FIBROSIS AT THE TIME OF REPAIR HAS LATE IMPACT ON THE DIASTOLIC FUNCTION AND ON THE INDEXES OF MYOCARDIAL DEFORMATION

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Introduction: We previously demonstrated that myocardial remodelling at the time of tetralogy of Fallot (TF) repair influences right ventricular (RV) function in the intermediate postoperative period. Its impact on the late postoperative RV function is unknown.
Objective: To re-evaluate previously studied patients and analyse RV function by echocardiography, correlated with the area of myocardial fibrosis at the time of repair.
Methods: 18 patients in the late follow-up (LFU) of TF repair (mean = 95 months, 13 males, age 12-80 months at repair and mean age at LFU = 126,6 months) were enrolled. Tissue-Doppler systolic and diastolic myocardial velocities (S', e', a') were determined: before surgery, 3 days postoperatively, between the 30th-90th PO days, at LFU (mean = 96.6months). Tricuspid annular plane systolic excursion (TAPSE) and RV peak longitudinal systolic global and regional strain (Strain) were analysed at LFU.
Results: The a' myocardial velocity on the RV lateral wall decreased significantly after surgery and persisted abnormal at LFU (RM ANOVA, p < 0.001). There was a significant negative correlation between e' velocity at LFU and myocardial fibrosis (p = 0.02; r = -0.54) and a positive correlation between fibrosis and E/e' ratio (r = 0.787; p = 0.0002). TAPSE was lower than the lower limit for normality in 16 patients. Global longitudinal RV systolic strain was decreased at LFU (<2std) in 14 patients and differed regionally in the mid septal segment (<2std, 5 patients) and in the mid segment of the lateral wall (<2std, 1 patient). There was negative correlation between myocardial fibrosis and systolic Strain in the mid septal segment (p = 0.0376; r = -0.493) but not with overall myocardial deformation index.
Conclusion: Myocardial fibrosis evaluated at the time of repair associated with late alterations in RV diastolic function and deformation index in the mid septal segment. Regional differences may correspond to adaptation of the RV components in the LFU and to the loading conditions. Fibrosis does not influence late systolic function.

O1526 - CONGENITAL HEART DEFECTS IN NIGERIAN CHILDREN PRELIMINARY DATA FROM THE NATIONAL PAEDIATRIC CARDIAC REGISTER

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Background/Hypothesis: Congenital heart defects (CHD) are the commonest birth defects and leading cause of birth defect related deaths. Incidence of significant CHD at birth is about 1% worldwide. However, there may be regional differences in pattern, even within countries. There has been no previous original study of regional patterns of CHD in Nigeria. Objective of the study is to compare current regional patterns of CHD and access to treatment in Nigerian children.
Materials and Methods: Seventeen centres nation-wide (6 from the north) participated in the pilot run of a National Paediatric Cardiac Register from January to December 2014. All children confirmed by echocardiography to have CHD were enrolled. Basic patient data from the Northern and Southern regions, such as biodata, echocardiographic diagnosis and details of any intervention

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received were compared with respect to pattern of disease and access to corrective surgery.
Results: A total of 1318 children including 701 (53.2%) males with CHD were enrolled during the one year pilot and 26.1% were from the North. Half of the patients were neonates and infants while almost one quarter had cyanotic defects. Median age at enrollment was 0.9 years (range 2 days to 18 years). Patients in the North were significantly older at enrollment (3.0 ± 3.7 years versus 2.3 ± 3.5years) – $p = 0.003$. Ventricular septal defects (VSD) were significantly more prevalent in the North (36.0% versus 20.6%) – $p < 0.0001$. Although 1265 (96%) of enrolled patients required surgical intervention, only 210 (16.6%) received the necessary surgery. Furthermore, only 43 (20.5%) of the surgeries done were in-country. More patients in the South (18.8%) received such intervention, compared with 10.5% in the North ($p < 0.0001$).
Conclusion: Over 80% of Nigerian children with CHD have no access to corrective surgery. There are significant differences in access to paediatric cardiology and cardiac surgical services between North and South of Nigeria.

O1860 - ECHOCARDIOGRAPHIC ACCURACY DETECTION OF CORONARY PATTERN IN D TRANSPOSITION OF THE GREAT ARTERIES A NEVER DESCRIBED NEW PATTERN IN OUR 10 YEARS' SINGLE CENTER EXPERIENCE

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Background: Assessment of coronary artery (CA) anatomy in transposition of the great arteries (TGA) is crucial for planning surgical technique. Echocardiography is still the main diagnostic tool for preoperative CA evaluation.
Aim: To review our last 10 years' TGA experience, and evaluate echocardiographic diagnostic accuracy in detection of CA pattern. Collaterally to determine coronary patterns prognostic implications.
Methods: We retrospectively reviewed echocardiographic and surgical records of TGA cases underwent primary repair in our institution from January 2004 to January 2014, comparing echocardiographic CA description to surgical findings.
Results: We collected 181 cases: 111 were TGA with intact ventricular septum (61%) and 70 TGA associated with ventricular septal defect (39%). Median age and weight at repair were 24 days (range 3-356 days) and 3,380 Kg (range 2,300-7,600 Kg) respectively. Coronary pattern were "usual" in 111 cases (61%) and were confirmed to be such in 95% (106/111). Abnormal coronary patterns were described in 70 patients (39%) and were confirmed in 87% of cases (61/70). Failure to detect coronary pattern was observed in 14 cases as described in the Table. Diagnostic accuracy for intramural CA was 75% but reached 100% in the last 6 years. We correctly identified by echocardiogram one never described coronary pattern: from right coronary ostium arose the right CA, the circumflex CA with posterior loop and an accessory anterior descending artery with anterior loop. From the left ostium arose the anterior descending artery. Global intra operative mortality was 0%; late mortality 1%.
Conclusion: Echocardiography is a very reliable tool in detecting CA in TGA even in complex patterns. Over the last 2 years intramural course detection improved, and we found a new, never described previously, CA pattern.

Table.

UNUSUAL CORONARY PATTERN	ECHOCARDIOGRAPHIC DIAGNOSIS	INTRAOPERATIVE DIAGNOSIS	N. cases	False Positive	False Negative
Circumflex artery	Cx from the RCA	Usual	4	4	2
	Usual	CX from the RCA	2		
Intramural arteries	Normal	Intramural LAD	1	1	1
	Intramural LAD	Normal	1		
Single coronary artery	Single coronary artery from right sinus	Usual	2	2	1
	Usual	Single coronary artery from left sinus	1		
Other	Single coronary artery arising from the left sinus	Inverted origin of the coronary arteries with anterior double loop of RCA and posterior loop of LAD	1	1	
	Usual	Separate origin of Cx and LAD	1		
	Single coronary artery from the left sinus	RCA arise from left sinus with anterior loop and Cx from right sinus with posterior loop	1		

LAD: left anterior descending coronary artery; RCA: right coronary artery; Cx: circumflex coronary artery

O1867 - ROUTINE CORONARY CT ANGIOGRAPHY IN YOUNG ADULTS WITH TRANSPOSITION OF THE GREAT ARTERIES AFTER THE ARTERIAL SWITCH OPERATION – IS THIS AN OPTION OR SHOULD IT BE MANDATORY

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Background: Coronary transplantation is the crucial part of successful arterial switch operation especially in the presence of the coronary anomalies. Coronary complications are relatively rare, but from the all postoperative adverse events they are potentially most dangerous. In the late follow-up those patients are frequently completely asymptomatic until a serious coronary event occurs. The Aim of This Study was to present the initial results of our routinely performed coronary CT angiographies (CCTA) in asymptomatic young adults with TGA after ASO.
Methods: This prospective study involves initial series of patients invited for the routine CCTA performed in asymptomatic young adults with TGA after ASO. In each of the case the evaluation of coronary arteries (CAs) included:
- the spatial setup of the great vessels,
- position and angulation of the coronary ostia with the aortic wall
- dimensions of the ostial and narrowest part of CAs,
- volume rendering 3D and curved, multiplanar reconstructions
Results: Among our group of patients with TGA after ASO (currently 715 patients) between September 2015 and October 2016 we performed 30 routine coronary CT angiographies (mean age at examination: 19.3 years). In the majority of the cases aorta was situated rightward and in front of the pulmonary trunk with median angle of great vessels to the sagittal plane of 220 (range: 3-48). The mean position angle for RCA was 460 (clockwise) and for LCA 400(counterclockwise). 12 patients had the acute coronary angulation (<300) of at least one coronary artery. The median height to width ratio for RCA ostium was 1.1; for LCA 1.2; however 2 patients had LCA H:W ratio which exceeds 2. In one patient we detected ostial narrowing caused by surrounding structures (pulmonary trunk), one patient had intramuscular

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course of LAD, and in two cases muscular band in segment 7 of LAD was present.

O2026 - RISK OF CARDIAC TACHYARRHYTHMIAS AND SUDDEN CARDIAC DEATH IN PATIENTS WITH REPAIRED TETRALOGY OF FALLOT A CARDIAC MRI BASED STUDY

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Background: Cardiac tachyarrhythmias are the leading cause of sudden cardiac death (SCD) in patients with repaired tetralogy of Fallot (TOF). We evaluated risk factors for SCD or sustained ventricular tachyarrhythmia (VT) and supraventricular tachycardia (SVT) in patients with repaired TOF.
Materials and Methods: Children and adults with repaired TOF who underwent cardiac magnetic resonance (CMR) imaging between 2007 and 2016 at four tertiary centers were retrospectively assessed. Potential risk markers, based on surgical history, CMR, electrocardiography and echocardiography, were analyzed for prediction of the primary composite endpoint of sustained VT/SCD, and for the secondary endpoint of SVT.
Results: A total of 531 cases were included. During a median follow-up of 2.6 (0.9-5.6) years, 29 (6%) reached the primary endpoint of VT/SCD and 51 (10%) the secondary endpoint of SVT. Multivariable cox proportional hazards regression identified right ventricular mass index (RVMi) ≥50 g/m2 (Hazard ratio [HR] 6.95; 95% confidence interval [CI] 3.14-15.4; $p < 0.001$), RV ejection fraction <45% (HR 2.93; 95% CI 1.30-6.60; $p = 0.009$), and QRS-duration ≥164ms (HR 4.81; 95% CI 2.17-10.7; $p < 0.001$) to be independent predictors of VT/SCD. Kaplan-Meier curve showed that in patients with all aforementioned risk factors, 69% experienced VT/SCD after nine years post-CMR [log rank $p < 0.001$]). Body mass index ≥25 kg/m2 (HR 3.33; 95% CI 1.79-6.16; $p < 0.001$) and older age at TOF repair ≥5 years (HR 2.74; 95% CI 1.47-5.12; $p = 0.002$) independently predicted SVT. Kaplan-Meier curve illustrated that in patients with both risk factors, 46% developed SVT within nine years following CMR [log rank $p < 0.001$]).
Conclusions: In patients with repaired TOF, RV hypertrophy, RV systolic dysfunction, and prolonged QRS duration were strong independent predictors of VT/SCD. Obesity and older age at TOF repair were associated with the occurrence of SVT. The present study therefore contributes to the risk stratification for sustained VT/SCD and SVT in patients with repaired TOF.

O2045 - IMPACT OF LOADING CONDITIONS ON VENTRICULAR FUNCTION BEFORE AND AFTER CONE RECONSTRUCTION IN EBSTEIN'S ANOMALY

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Background/Hypothesis: The Cone reconstruction in Ebstein's anomaly (EA) aims to reduce tricuspid valve regurgitation (TR) and reposition the valve to the anatomic annulus, but post-operative progress of ventricular function is poorly understood. This study evaluated the ventricular adaptation to the altered loading conditions after Cone reconstruction.
Material and Methods: A retrospective study assessing longitudinal change was conducted from 2009 to 2014. All symptomatic patients with EA and severe TR undergoing surgery were included. Transthoracic advanced echocardiography was performed pre- and postoperatively (at short-term (<30 days) and mid-term). Conventional and longitudinal 2D strain parameters were measured for LV and RV. Paired analyses were compared using Wilcoxon Matched-pairs signed rank test.
Results: The echocardiographic data of 17 patients, aged 15 (1-57 years) at operation could be analysed. Median follow up was 6 months (8 days-54 months). GOSH score was significantly reduced after Cone repair (1.1 ± 0.2 vs 0.3 ± 0.1, $p = 0.020$) as well as TR (4 (3-4) vs 1 (0-3), $p < 0.001$). The TAPSE (26.4 ± 5.8mm vs 8.7 ± 3.2mm, $p < 0.001$), RV FAC ($45.0 \pm 8.1\%$ vs $35.5 \pm 5.8\%$,$p = 0.038$) and LV 2D peak systolic strain were significantly reduced postoperatively (-20.5 ± 2.8 vs -17.7 ± 2.8, $p = 0.041$), but the LV 2D strain tended to recovery in mid-term postoperative period (-20.7 ± 3.1 in pre-op vs 18.8 ± 2.4 in mid-term post-op, $p = 0.247$). There was no evidence of systolic mechanical dyssynchrony.
Conclusions: Although clinical outcome of Cone reconstruction remains excellent, acute post-operative changes in loading conditions leads to reduction of myocardial contractility of both ventricles, with a trend to later recovery for LV.

O2073 - THREE DIMENSIONAL ECHOCARDIOGRAPHY VERSUS CARDIAC MAGNETIC RESONANCE FOR ASSESSMENT OF THE RIGHT VENTRICULAR VOLUMES IN PATIENTS WITH CONGENITAL HEART DISEASE AFTER PERCUTANEOUS PULMONARY VALVE IMPLANTATION

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Background: Cardiac MR (CMR) is the gold standard for right ventricular (RV) quantification. In the recent years transthoracic three-dimensional echocardiography (3DE) is becoming increasingly available in the routine clinical practice. The purpose of this study was to investigate the clinical significance and interchangeability of these modalities to evaluate patients with congenital heart disease (CHD) who underwent percutaneous pulmonary valve implantation (PPVI) for RV outflow tract dysfunction.
Materials and Methods: 52 patients who underwent PPVI were evaluated with 3DE and CMR to quantify the RV. RV volumes and ejection fraction (EF) were measured for both imaging techniques with commercially available softwares (Tomtec-Germany for 3DE and Medimatic-Netherlands for CMR data). Paired t test,

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Bland-Altman analysis and Pearson's correlation analysis were used as most appropriate.
Results: 80% of the patients had adequate image quality on 3DE and was included in the study. 78% had a primary disease of the RV (tetralogy of Fallot, pulmonary atresia, transposition of the great arteries,…), while 9 patients had a natively normal RV and underwent PPVI after a Ross procedure. No differences were found regarding age, body size and type of valve implanted. Linear regression analysis showed high correlation coefficients between 3DE and CMR ($p < 0.001$). Bland-Altman analysis showed limited bias and narrow limits of agreement (LOA) between the two techniques: End Diastolic Volume index (EDVi): bias -2.1 ml, LOA ±22.8 ml; End Systolic Volume index (ESVi): bias -4.1 ml, LOA ±15 ml; EF: bias 4%, LOA: ±16.3. A non-significant trend towards overestimation of volumes by 3DE was observed. The 3DE measurements were found to be highly reproducible in terms of intra-observer variability (EDVi 0.97, ESVi, 0.87, EF 089, $p < 0.001$).
Conclusions: The present study supports the applicability of 3DE for the assessment of RV volumes and EF in patients with CHD after PPVI.

Table 1. Comparison of RV volumes and EF between 3DE and CMR

	CMR	3DE	P*	r
EDVi (ml/m^2)	82.98 ± 23.52	85.12 ± 22.48	0.001	0.873
ESVi (ml/m^2)	37.33 ± 17.60	41.4 ± 15.60	0.001	0.897
EF (%)	56 ± 10.6	52.04 ± 9.89	0.001	0.675

P value for % difference

O2162 - MULTI PARAMETRIC ECHOCARDIOGRAPHIC ASSESSMENT FOR PREOPERATIVE DECISION MAKING IN SURGICAL TREATMENT OF ATRIOVENTRICULAR SEPTAL DEFECT

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Introduction: Surgical management of unbalanced atrioventricular septal defect (AVSD) remains challenging because of a high mortality rate and difficulties in decision making between biventricular repair (BVR) and univentricular palliation (UVP) in borderline cases. In this study we aimed to define echocardiographic criteria for imbalance and prove their predictability for mortality and reoperation rate.
Methods: From 1986 to 2016 671 patients diagnosed with AVSD were identified in our institution. 532 patients received AVSD repair, 2.5% of these patients had unbalanced AVSD but were suitable for biventricular strategy. Of 253 patients pre- and postoperative echocardiographic data were available for retrospective analysis. For echocardiographic assessment modified atrioventricular valve index (mAVVI), ventricular cavity ratio (VCR), left ventricular inflow index (LVII) and right ventricle/ left ventricle inflow angle were measured. Left atrioventricular valve reduction index (LAVRI) was calculated to estimate left atrioventricular valve area after cleft closure. Correlation was proved between echocardiographic indices, surgical strategy, postoperative course, survival and reoperation rate.
Results: mAVVI was the only index to distinguish between balanced and unbalanced AVSD and predict surgical strategy (Jegatheeswaren et al.). Borderline cases (n = 13) received biventricular repair and had no higher mortality or reoperation rate. Inflow angle positively correlated with ventilation time ($p < 0.01$) and predicted early postoperative course. LAVRI <1.0, moderate mitral valve regurgitation and a mean gradient >3 mmHg on discharge were associated with reoperation.
Conclusion: No index correlated with mortality. mAVVI seemed usable for preoperative decisions concerning interventricular balance. Correlation of right ventricle/left ventricle inflow angle with ventilation time may indicate the importance of ventricular septal defect size for postoperative course. LAVRI and residual mitral valve regurgitation/stenosis correlated with reoperation rate. A multi-parametric echocardiographic assessment using indices for ventricular and atrioventricular valve size can facilitate decision making in borderline cases of unbalanced AVSD and surgical management of the atrioventricular valve.

O2224 - ROLES OF PRESSURE AND VOLUME LOAD ON NATURAL PROGRESSION OF RIGHT VENTRICULAR DILATION AND REVERSE REMODELING AFTER PULMONARY VALVE REPLACEMENT IN PATIENTS WITH REPAIRED TETRALOGY OF FALLOT A SERIAL FOLLOW UP CARDIOVASCULAR MAGNETIC RESONANCE STUDY

Chun-An Chen[1], Shyh-Jye Chen[2], Wen-Yih Tseng[3], Yao-Hui Tseng[2], Mao-Yuan Su[2], Mei-Hwan Wu[1], Chun-Wei Lu[1], Ming-Tai Lin[1]
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Background: Different patterns of right ventricular (RV) remodeling in response to different types of hemodynamic burden have been observed in patients with repaired tetralogy of Fallot (rTOF). However, little is known about the impact of hemodynamic load on the progression rate of RV dilation, nor on the RV reverse remodeling after pulmonary valve replacement (PVR). We sought to investigate these issues using serial CMR studies.
Methods: We identified 188 rTOF patients who had received at least 2 CMR studies from our institutional database (482 CMR studies). The natural progression rate of the RV end-diastolic volume index (EDVi) (RVEDV PR) was calculated using the change in the RV EDVi over time. Patients with a pulmonary regurgitation (PR) fraction ≥ 25% and peak RV outflow tract gradient of ≤25 mm Hg were classified into RV volume overload group, and all the remaining patients were classified into mixed load group.
Results: Over a median interval of 3.9 years, the median RVEDV PR was 4.6 mL/m2/y (n = 169). Patients with upper-quartile RVEDV PR (>7.5 mL/m2/y) were younger at the time of first CMR study and repair; had a higher baseline PR fraction, larger RV dimensions, and a lower left ventricular ejection fraction; and were more likely to have volume overload as the predominant hemodynamic burden (n = 91) as oppose to mixed load (n = 78). In multivariate analysis, predominant RV volume overload was independently associated with upper-quartile RVEDV PR. Forty-one patients subsequently received PVR, and RV size decreased substantially. However, it was pre-PVR RV size, age at

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PVR, and post-PVR PR fraction independently associated post-PVR RV size, but not types of hemodynamic burden before PVR.
Conclusions: Patients with predominant RV volume overload are at a higher risk of rapid natural progression of RV dilation. However types of hemodynamic burden before PVR was not related to the extent of RV reverse remodeling after PVR.

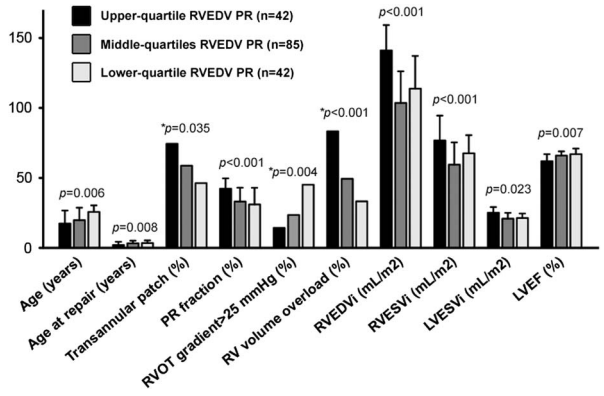

Figure.

O2255 - QUANTIFICATION OF PULMONARY VALVE REGURGITATION IN PATIENTS WITH TETRALOGY OF FALLOT BY A NOVEL ECHOCARDIOGRAPHIC IMAGING OF VECTOR FLOW MAPPING

Satoshi Yasukochi[1], *Junpei Kawamura*[1], *Yusuke Nakano*[2], *Kiyohiro Takigiku*[1], *Kouta Takei*[1], *Sei-ichi Tazawa*[1], *Masashi Utsumi*[1]
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Background: The pulmonary valve regurgitation (PR) is a key to determine a long-term outcome after surgical treatment of Tetralogy of Fallot (TOF) by causing right ventricular (RV) dilatation and dysfunction. Only quantification for PR could be measured by cardiac magnetic resonance (CMR).
Aim: We aim to quantify PR of patients with TOF by a novel imaging modality of echocardiography: Vector Flow Mapping (VFM) and to validate the results of this new imaging by CMR-Flow study.
Method: Thirty-four patients after surgical repair of TOF, mean age at examination of 14.8 years and interval from definitive surgery of 11.9 years, were investigated. The color Doppler images of RV outflow tract including main pulmonary artery was obtained by Prosound F75 (Hitachi-ALOKA ltd) with VFM mode, and calculated the blood volume across the measured line perpendicular to pulmonary valve ring (PV) as a sum of integration of blood flow velocity at multiple sampling gate along a cardiac cycle by off-line analyzing software (Hitachi-Aloka ltd.) The ratio of regurgitation to forward flow at PV measured by VFM (PRF-VFM) was compared to those by CMR which were measured by phase-contrast flow quantification (PRF-CMR).
Results: PRF-VFM was good in agreement with PRF-CMR: (PRF-CMR) = 0.917*(PRF-VFM), $r^2 = 0.63$ as Figure 1. Limitation: The limitation of this technique is the quality of image of RVOT and a color Doppler signals, especially in the presence of turbulent accelerated flow by stenotic RVOT because of aliasing artifacts.
In conclusion, our new imaging modality of VFM is very powerful tool to determine PRF in as accurate quantification as that of CMR when to diagnose the severity of PR in patients with TOF.

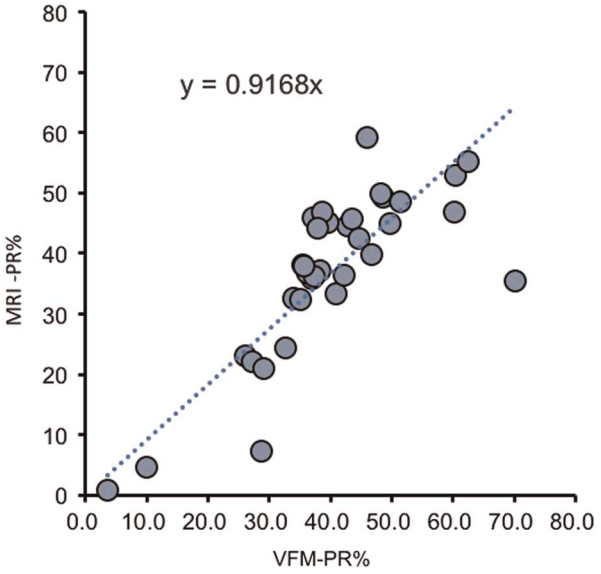

Figure 1.

O2309 - MYOCARDIAL CONTRACTILE RESERVE AND CORONARY FLOW RESERVE FOLLOWING ARTERIAL SWITCH OPERATION FOR TGA REGRESSED AND PRESERVED LV

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Introduction: The functional capability of the 'regressed' left ventricle (LV) with transposition of great arteries (TGA) may be different from those who underwent early arterial switch operation (ASO). The functional reserve and coronary flow reserve may also differ and have never been studied before.
Methods: All patients who underwent an ASO for TGA at least 6 months prior were included in the study. All the patients underwent a detailed echocardiography followed by adenosine stress echocardiography.
Results: We included 40 patients for echocardiography, with 22 patients (mean age 276.45 ± 121 days) undergoing an ASO with a preserved LV and 18 patients (mean age 336.17 ± 115.88 days) with a regressed ventricle at time of surgery. The LV ejection fraction was within normal limits at baseline (63.19 ± 10.95 Vs 60.44 ± 14.24; P value 0.50) and increased with adenosine stress in both the groups (67.31 ± 8.71 Vs 61.58 ± 13.11; P value 0.12), however, the increase was not statistically significant. Similarly, there was no significant difference in the parameters including longitudinal, circumfential and rotational strain between both the groups. The change following adenosine stress was also not significant in both the groups. However, the coronary flow reserve seems to be preserved.
Conclusion: Patient with regressed left ventricle undergoing arterial switch operation adapt well to systemic afterload and perform as well as patients of arterial switch operation with preserved left ventricle at 6 months follow up. The myocardial contractile reserve seems to be reduced in both the groups despite a preserved coronary flow reserve.

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O2531 - CARDIAC OUTPUT DISTRIBUTION IN THE FETUS WITH COARCTATION

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Introduction: Coarctation of the aorta (COA) remains one of the most challenging diagnoses by fetal echo, with false positive diagnosis being quite common. Although relative left-sided anatomic hypoplasia is well described in these fetuses, the physiological aberrations have not been well characterized. In the normal fetus, the left ventricle supplies approximately 40% of the combined ventricular output (CVO) but it is unknown whether this holds true in fetuses with COA. We hypothesized that patients with confirmed neonatal COA would have lower left ventricular (LV) output in utero.
Methods: We included 41 patients who underwent surgery for neonatal COA between 2005 and 2015 and analyzed data from their fetal echos, including aortic and pulmonary valve size (AOV and PV) and ventricular output. Findings were compared to our institutional normal fetal data, matched for gestational age (n = 177).
Results: Median gestational age was identical in both cohorts (24.71, p = 0.8).The AOV z-score was significantly lower in the COA cohort (-2.2 vs -0.155, p < 0.0001), and the PV z-score higher (0.43 vs -0.012, p = 0.0071). The LV component of CVO was lower in COA than in controls (median of 28.9% vs 41.3%, p < 0.0001), as was the total CVO (449 ml/kg/min vs 553 ml/kg/min, p = 0.0012). Both the AOV annulus (p = 0.005) and the AOV Doppler velocity-time integral (VTI) (p < 0.0001) increased less with advancing gestational age in patients with COA than in normal fetuses.
Conclusion: Fetuses with confirmed neonatal COA have significantly reduced LV component of CVO. This is likely due to a smaller AOV as well as reduced VTI. The reduced VTI may in turn be a result of reduced preload from lower amount of right-to-left flow at the foramen ovale. Further work is necessary to elucidate this pathophysiology and evaluate its potential for enhancing prenatal diagnosis of COA.

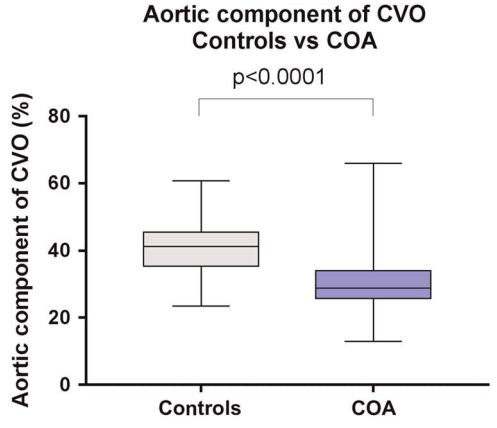

Figure.

O2664 - DIASTOLIC KINETIC ENERGY MEASURED BY 4D FLOW MAGNETIC RESONANCE IMAGING IS RELATED TO RESTRICTIVE RIGHT VENTRICULAR PHYSIOLOGY IN PATIENTS WITH REPAIRED TETRALOGY OF FALLOT

Pia Sjöberg[1], Sebastian Bidhult[1], Jelena Bock[1], Einar Heiberg[1], Håkan Arheden[1], Shahab Nozohoor[2], Ronny Gustafsson[2], Marcus Carlsson[1]
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Background: The clinical relevance of restrictive right ventricular (RV) physiology in patients with pulmonary regurgitation (PR) after repair of Tetralogy of Fallot (rToF) is debated. 4D-flow cardiovascular magnetic resonance (CMR) enables quantification of ventricular kinetic energy (KE) which may provide improved understanding of the pathophysiology. The aim of the study was to quantify ventricular KE in patients with PR after rToF and evaluate the impact of restrictive RV physiology.
Materials and Methods: Fifteen patients with rToF and PR (5 females, median age 24 years, range 18-52) underwent CMR including four-dimensional (4D) phase-contrast flow sequence. Thirteen healthy volunteers (2 females, median age 28, range 23-43) were used as controls. Ventricular segmentation was performed in cine SSFP images and imported to 4D-flow dataset. KE in the RV was calculated as $KE = \frac{1}{2}mv^2$ for each time frame. Restrictive physiology was defined as forward flow in the pulmonary artery during atrial contraction measured by 2D-flow CMR. Results are presented as mean ± SD.
Results: No difference was seen in RV peak systolic KE in patients, 10.2 ± 4.2mJ compared to controls, 8.3 ± 2.0 mJ, p = 0.21. RV peak diastolic KE was higher in patients, 8.0 ± 5.1mJ than controls, 3.1 ± 1.3mJ, p = 0.0002, also indexed to stroke volume (SV), 0.06 ± 0.03mJ vs 0.03 ± 0.01mJ, p = 0.0004. Examples are shown in fig 1. Mean peak systolic KE/peak diastolic KE ratio was lower, 1.6 ± 0.9mJ, p = 0.0004, in patients versus controls, 2.9 ± 1.1mJ, fig 2. Patients with a nonrestrictive physiology (n = 5, open circles) had ratio < 1 or in one case just above 1, whereas ratio > 1 was related to restrictive RV (n = 10, filled circles). Cohen's kappa 0.84.
Conclusions: Peak diastolic KE is increased in rTOF with PR, but the flow pattern in restrictive rTOF is more similar to controls and further studies are necessary to evaluate the impact on clinical outcome.

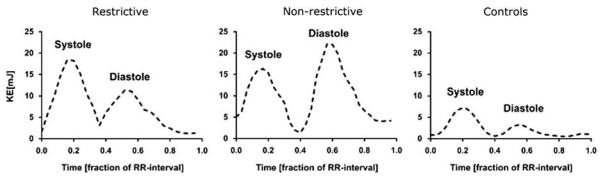

Figure 1.

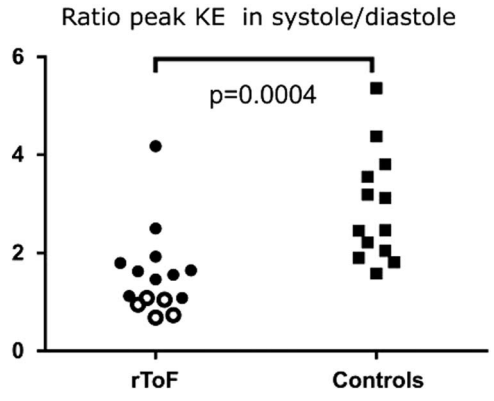

Figure 2.

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O2835 - TRICUSPID VALVE ROTATION ANGLE (TVRA) PREDICTS THE SUCCESS OF CONE TYPE RECONSTRUCTION FOR EBSTEIN'S ANOMALY; NOVEL APPROACH USING CARDIAC MRI

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Aim: To identify critical cardiac MR-derived attributes of the variable Ebsteinoid tricuspid valve (TV) that could preclude effective cone-type surgical reconstruction.
Methods: A retrospective assessment of all adult and pediatric patients with Ebstein's anomaly of TV and pre-operative CMR, undergoing cone-type surgical reconstruction, by a single surgeon, within two surgical centres. CMR-derived geometric measurement of the displacement and rotation of the plane of systolic apposition of the leaflets (TVRA angle), in the RV vertical long axis image, was related to the short-term surgical result of all operated patients. The primary outcome measure was early dehiscence, re-operation or modified surgical plan. Other CMR factors assessed: area ratios of cardiac chambers (from 4-chamber view), BSA-indexed functional RV volume (RVEDVi), and TV regurgitant fraction (TR%). Non-parametric statistical tests compared the groups.
Results: Since the first procedure in 2010, 33 patients (15 male) had pre-op CMR and underwent surgery at a median (25th-75th C) age 16 (3-57) years. 5/33 patients experienced early surgical failure, with even temporal spread between 2010-2016. Significant risk factors for technical failure of repair were increased TVRA (median 73degrees (62-78) vs 49 degrees (39-60), $p < 0.01$), smaller RVEDVi (98 mL/m^2 (66-141) vs 173 mL/m^2 (121-212), $p = 0.02$), decreased TR% (36% (23-53) vs 59% (43-70), $p = 0.046$) and increased age (35 yrs (19-49) vs 15 yrs (8-21), $p = 0.04$. Other CMR indicators of Ebstein disease severity: 'RA area: rest of heart' ratio, 'Right heart:Left heart' area ratio, cardiothoracic ratio or RV ejection fraction did not relate to early surgical failure. The TVRA was measureable even with poor image quality and showed low inter-observer variability.
Conclusions: A measured angle of rotation of the effective plane of closure of the Ebsteinoid TV may be a simple, reliable marker of the technical feasibility of cone-type surgical reconstruction. Though prospective testing is needed, this angle may improve case-selection and therefore surgical outcome.

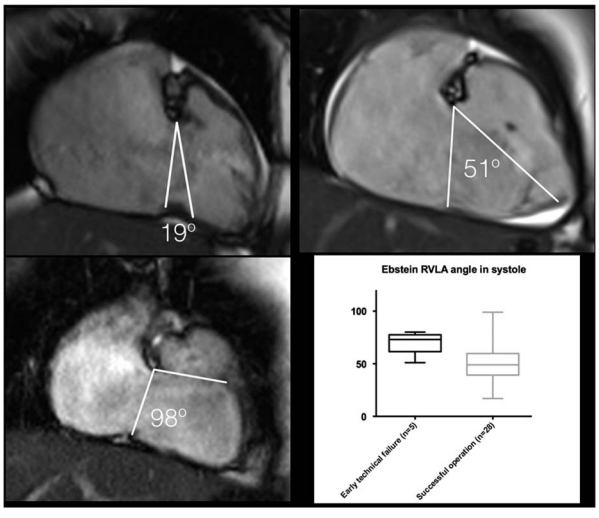

Figure.

INTENSIVE CARE

O1145 - RISK STRATIFICATION OF PREMATURE BABIES WITH CONGENITAL CARDIAC DISEASE

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Objectives: Describe a risk stratification for prematurity associated with congenital heart disease (CHD).
Methods: 11 years retrospective case control review of a neonatal database "BadgerNet". Outcome measures of 154 premature babies with congenital cardiac disease were compared with pre-term controls (175) without congenital cardiac abnormalities. We enrolled all preterm babies with CHD born before 36 gestation weeks at St Thomas' Hospital. Babies with isolated patent ductus arteriosus were excluded to avoid bias. In order to describe the risk stratification we randomly selected a control group of premature babies without CHD born before 36 gestation weeks. Among the two groups birth weight, gender and gestational age was similar.
Results: Our primary outcome was survival to discharge from neonatal unit. Of 154 live born premature (below 36 weeks gestation) infants between January 2005-December 2015, (12.33%) died in neonatal intensive care compared to controls (4%). The odds ratio(OR) of dying in prematurity with CHD was 3.37(95% confidence interval [CI]: 3-3.74). The incidence of necrotizing enterocolitis (NEC) among our study group was (16.88%) versus controls (7.42%)giving odds ratio of 2.53(95% confidence interval [CI]: 2.16-2.9). We also looked at sepsis and (66.88%) premature babies with CHD were treated for it compared to (55.42%) controls, odds ratio (OR) 1.62(95% CI: 1.32-2.06).The mean length of stay in neonatal unit was also significantly longer: 36.94 days versus 24.25 days for controls. We divided our population into four groups: 23-26 gestation weeks (GW), 27-29 (GW), 30-32 (GW) and 33-36 (GW) and the most common cardiac diagnostic among the 23-26 (GW)premature babies was pulmonary stenosis/atresia (40%) while in the last category (22.05%) had coarctation of the aorta followed by (13.23%) transposition of great arteries and truncus arteriosus.

O1152 - PRE OPERATIVE NUTRITION IN NEONATES UNDERGOING SURGERY FOR CONGENITAL HEART DISEASE AND ITS IMPACT ON POSTOPERATIVE COURSE

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Background: Nutrition is vital in maintaining optimal cellular and organ function, particularly in neonates undergoing congenital heart surgery. Achieving nutritional goals preoperatively is challenging, particularly with fluid restrictions and suboptimal oral intake. We sought to evaluate our ability to achieve pre-operative caloric goal and ascertain its effects postoperatively.
Materials and Methods: We retrospectively reviewed neonates from 2011-2015 who underwent congenital heart surgery at Arnold Palmer Hospital for Children. Multiple nutritional and post-operative variables were collected. Study outcomes included hospital length of stay (HLOS) and duration of mechanical

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ventilation (DMV). T test, chi-square, and multivariate analysis were used.
Results: 121 neonates were reviewed. 79 (65%) patients did not achieve preoperative caloric goal with a (median age 5 days), while 42 (35%) did (median age 13 days). Of those achieving caloric goal, 35 (83%) achieved it via total parental nutrition (TPN) or a combination of TPN and enteral feeds. 85 patients (70%) achieved their protein goal. Catheterization/reoperation ($p = 0.03$) and high-grade acute kidney injury (AKI) ($p = 0.008$) were more likely to occur in those not achieving caloric goal preoperatively. Those with higher STAT category ($p = 0.01$) were less likely to achieve caloric goal. Those not achieving caloric goal had a longer hospital length of stay (HLOS) ($p = 0.02$). There was no statistically significant difference in the incidence of sternal wound infections. Multivariable analysis with interaction term using AKI as the dependent variable found that failure to achieve caloric goal preoperatively increased the adverse effect of lower protein intake on renal function and was associated with AKI (0.04).
Conclusion: Achieving preoperative nutritional goal in neonates undergoing congenital heart surgery is challenging. Failure to achieve calorie and protein goals may lead to increased risk of high-grade AKI and prolonged HLOS.

O1187 - EFFECT OF GUIDELINES BASED MANAGEMENT ON THE EARLY POST OPERATIVE OUTCOME AFTER BLALOCK TAUSING SHUNT

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National Guard Hospital, Pediatric Cardiac Intensive Care, Riyadh-Saudi Arabia[1]; National Guard Hospital, Ational and Gulf Center For Evidence Based Health Practice, Riyadh-Saudi Arabia[2]

Background and Aim: Modified Blalock- Tausing shunt (MBTS) is a palliative procedure in cyanotic heart diseases to facilitate blood flow to the lung (1), though it showed significant post-operative mortality and morbidity especially in the neonatal group. to master the post-operative management and minimize complication, mortality and morbidity, we designed and implemented executive guidelines to maintain patients respiratory and hemodynamically stable especially during the critical early post-operative period. Aim of our study to evaluate the patient outcome after implementation of these guideline in our center.
Methods: We conducted a retrospective chart review analysis of all children who underwent MBTS since year 2000 till December 2015, we excluded cases of hypoplastic left heart syndrome. We compared the early outcome of patients operated after the guidelines implementation (March 2013 till December 2015) (group A) with patients operated before implementing the MBTS guidelines (group B). Also we compared the early outcome of patients showing over-circulation (over-shunting) with other patients after implementation of the guidelines.
Results: 197 patients underwent MBTS since year 2000 till December 2015, 25 patients (64% neonates) after the implementation the guidelines (group A), and 172 patients (60% neonates) before the guidelines implementation (group B). there was a significant improvement in the postoperative course and morbidity after guidelines implementation. The ventilation time, reintubation rate, inotropic support duration, and postoperative complications were significantly lower in (group A), there was a trend of higher mortality in group B (15.7% VS 8%) (p 0.5), but it was more obvious in the neonatal patients group (19.41% VS 6.25) (p 0.2).
Conclusion: This study prove that protocol based management of patients after MBTS can improve the postoperative course and outcome.

O1275 - HETEROTAXY SYNDROME AND INTESTINAL ROTATION ABNORMALITIES – A PROSPECTIVE STUDY

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Stollery Children, Pediatric Cardiac Intensive Care Unit, Edmonton-Canada[1]; University of Alberta, Department of Pediatrics, Edmonton-Canada[2]; University of Saskatchewan, Department of Pediatrics, Saskatoon-Canada[3]; University of Manitoba, Department of Pediatrics, Winnipeg-Canada[4]; University of Calgary, Department of Pediatrics, Calgary-Canada[5]; Nicklaus Children, Division of Cardiovascular Surgery, Miami-United States[6]; Advocate Children, Advocate Children, Oak Lawn-United States[7]; University of Nebraska Medical Center, Division of Pediatric Surgery, Omaha-United States[8]; University of Alberta, Department of Surgery, Edmonton-Canada[9]

Background: Infants with heterotaxy syndrome (HS) have an abnormal arrangement of organs along the right-left body axis. Malrotation is the most clinically significant intestinal rotation abnormality (IRA). The objective of this study was to prospectively observe a cohort of infants with HS and determine the incidence and natural history of IRA and volvulus in this population.
Materials and Methods: Prospective observational study; inclusion criteria were a diagnosis of HS and age ≤ six months. HS was defined as any arrangement of organs that was not complete situs solitus or situs inversus with associated congenital heart disease. Screening for IRA was at the discretion of each participating center. IRA was classified as positive, negative, indeterminate or nonrotation.
Results: Subjects were recruited from January 2012 to December 2016. Forty infants from seven institutions across North America were included; 22 subjects had right atrial isomerism and 18 had left atrial isomerism. Median length of follow up was 587 days (range 21-1823). 31/40 infants (78%) were screened, all with upper gastrointestinal imaging. Ten were screened because of symptoms. 23 infants (74%) had IRA; malrotation ($n = 14$), nonrotation ($n = 7$), and indeterminate ($n = 2$). Both patients with indeterminate rotation had duodenal atresia. Of the ten patients screened due to symptoms, nine had IRA and two infants had an emergent Ladd procedure. Of the 30 infants either not screened or screened routinely, two (6.7%) had emergent laparotomy and Ladd procedure for symptoms they developed at 18 and 20 days. Only one infant had a prophylactic Ladd procedure. Of the four symptomatic infants who had an emergent Ladd procedure, none had volvulus. No infant presented later than two months of age with symptoms of IRA.
Conclusions: IRA is common in HS patients, most commonly malrotation. Only 10% of the cohort had an emergent Ladd procedure; volvulus was not observed.

O1311 - RISK FACTORS FOR AND BURDEN OF ACUTE KIDNEY INJURY IN CHILDREN UNDERGOING CARDIAC SURGERY FOR CONGENITAL HEART DISEASE

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Background: Acute kidney injury (AKI) is common after pediatric cardiac surgery (CS), which is associated with morbidity and mortality. There are sparse data regarding associations with specific lesions and cost.
Materials and Methods: Pediatric admissions with CS were identified in the Kids Inpatient Database (KIDS). The presence of AKI defined by ICD-9 codes during the included admissions was then identified. Regression analysis was done to determine factors associated with AKI and factors associated with longer length of stay, cost of stay, and greater need for extracorporeal membrane oxygenation (ECMO).
Results: 60,216 admissions were included in this analysis; 1,708 (3%) were documented to have AKI. AKI admissions were associated with: younger patient age (2.0 vs 3.1y, $p < 0.001$), longer stay (50 vs. 13 days, $p < 0.001$), more ECMO (20.6% vs. 1.3%, $p < 0.001$), increased cost ($620,156 vs. $153,596, $p < 0.001$) and increased mortality (31.8% versus 2.1%, $p < 0.001$). Cardiac lesions most associated with AKI were total anomalous pulmonary venous connection (OR 3.1, 2.6-3.7; $p < 0.001$), hypoplastic left heart syndrome (OR 2.4, 2.1-2.7; $p < 0.001$) and common arterial trunk (OR 2.2, 1.8-2.8; $p < 0.001$). CS most associated with AKI were heart transplant (OR 21.6, 19.2-24.2; $p < 0.001$), Fontan (OR 8.4, 6.9-10.2; $p < 0.001$), and complete repair of common arterial trunk (OR 3.2, 2.5-4.1; $p < 0.001$).
Conclusions: While AKI incidence was low compared to published reports, likely due to undercoding, we observed that AKI significantly increases mortality, length, and cost of stay. The postoperative AKI risk observed with underlying cardiac lesion and CS provides potential guidance for increased AKI surveillance and targeted AKI interventions in these populations.

O1359 - DEVELOPMENT AND IMPLEMENTATION OF INPATIENT GOAL DRIVEN CLINICAL PATHWAYS FOR CONGENITAL HEART SURGERY PATIENTS

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Background: With advances in the fields of congenital cardiac surgery and cardiac intensive care resulting in improved outcomes and survival, there is increased interest in decreasing morbidity and length of stay to better optimize long-term outcomes. Longer length of stay following cardiac surgery is associated with increased risk of infection, poor neurodevelopmental outcomes, and increased hospital costs. To these ends, development of simplified, "user-friendly" evidence informed, clinical practice pathways is paramount.
Objectives: To design and implement goal driven standardized clinical pathways for cardiology inpatient teams at Cincinnati Children's Hospital Medical Center, in an effort to reduce practice variation and promote evidence-based medicine.
Methods: A multidisciplinary team designed goal driven standardized clinical pathways for nine STS Benchmark postoperative cardiac surgical procedures. The three goals for all clinical pathways were: expedite extubation time, and reduce CICU and hospital length of stay. Each pathway recommends specific interventions to accomplish the goals. The team employed the Improvement Science Model and used Plan-Do-Study-Act cycles to refine each clinical pathway, identify practice variation, and address communication barriers. Balancing and outcome measures were calculated using statistical process control methodology.
Results: In the 20-month period following implementation, 131 patients were enrolled onto a clinical pathway. 120 patients met the recommended duration of mechanical ventilation goal for a 92% success rate. 92 patients met the recommended CICU length of stay goal for a 70% success rate. 83 patients met the recommended hospital length of stay goal for a 63% success rate. The balancing measures of ward to CICU bounce-back rate was low at 4% and the 30 day readmission rate of all cause was only 2%.
Conclusions: Implementation of goal driven clinical pathways revealed key lessons regarding practice variation, communication failures and unexpected patient clinical status. Duration of mechanical ventilation and length of CICU stay has improved.

O1458 - MULTICENTER VALIDATION OF THE VASOACTIVE VENTILATION RENAL SCORE AS A PREDICTOR OF PROLONGED MECHANICAL VENTILATION AFTER NEONATAL CARDIAC SURGERY

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Objectives: We sought to validate the vasoactive-ventilation-renal score (VVR), a novel disease severity index, as a predictor of prolonged duration of mechanical ventilation (DMV) in a multicenter cohort of neonates who underwent cardiac surgery.
Methods: Neonates less than 30 days old who underwent cardiac surgery at seven institutions within the United States in 2015 were retrospectively reviewed. VVR is calculated as follows: VVR = ventilation index + vasoactive-inotrope score (VIS) + Δ creatinine [change in serum creatinine from baseline*10]. Prolonged DMV was defined as greater than 96 hours (ICD-10-CM procedure code 5A1955Z). Receiver operative characteristic curves were generated and the abilities of the VVR obtained at three time points - ICU admission and 6 and 12 hours postoperatively - and the peak value of these three measurements to correctly classify patients as having prolonged DMV were compared to other predictor variables using area under the curve (AUC) values. Multivariable logistic regression modelling was also performed.
Results: We reviewed 273 neonates. Median age at time of surgery was 7 days (interquartile range: 5-12 days). Eight-six (31%) had single ventricle anatomy. Prolonged DMV occurred in 89 patients (32%). AUC values for the VVR at each time point as predictors of prolonged DMV were significantly greater than values for the corresponding ventilation index, VIS, and lactate (Table). The VVR also outperformed STAT mortality category as a predictor of prolonged DMV (Table). On multivariable regression analysis, after adjustment for potential confounders, VVR measurements at each time point were independent predictors of prolonged DMV (Odds ratio for peak VVR: 1.08, 95% confidence interval: 1.04, 1.12).
Conclusions: In a multicenter cohort of neonates who underwent cardiac surgery, the VVR was a reliable predictor of postoperative DMV and outperformed more traditional measures of illness severity.

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Table. Relationship between Predictor Variables and Prolonged Duration of Ventilation.

Predictor Variables	Median(IQR)	Maximum	AUC (95% CI)
STAT Mortality Category	4 (3-5)	5	0.66 (0.59-0.72)
Admission lactate (mg/dL)	3 (1.7-5)	13.1	0.68 (0.61-0.75)
6-hr lactate (mg/dL)	2.7 (1.6-4.1)	13.4	0.68 (0.61-0.75)
12-hr lactate (mg/dL)	2.2 (1.5-3.2)	11.6	0.66 (0.59-0.73)
Peak lactate (mg/dL)	3.7 (2.3-5.3)	13.4	0.69 (0.62-0.76)
Admission VIS	8 (5-11)	30	0.69 (0.62-0.76)
6-hr VIS	8 (5-11)	37	0.68 (0.61-0.75)
12-hr VIS	8 (5-12)	64	0.69 (0.62-0.75)
Peak VIS	10 (6-13)	64	0.70 (0.63-0.77)
Admission ventilation index	19 (15-25)	112	0.72 (0.66-0.79)
6-hr ventilation index	17 (13-22)	96	0.76 (0.70-0.83)
12-hr ventilation index	16 (12-21)	74	0.74 (0.67-0.80)
Peak ventilation index	21 (16-27)	112	0.78 (0.71-0.84)
Admission VVR	28 (22-38)	127	0.78 (0.73-0.84)
6-hr VVR	27 (21-34)	109	0.81 (0.75-0.86)
12-hr VVR	26 (20-33)	92	0.82 (0.76-0.87)
Peak VVR	31 (24-40)	127	0.82 (0.77-0.88)

[a]AUC: Area under the receiver operating characteristic curve; CI: confidence interval; IQR: interquartile range; STAT: Society of Thoracic Surgeons-European Association for Cardio-Thoracic
Surgery Congenital Heart Surgery mortality category; VIS: vasoactive inotrope score; VVR: vasoactive-ventilation-renal score.

[b]Patients extubated between 0 and 6 hours postoperatively (n = 4) were excluded from 6-hour and 12-hour analyses; patients extubated between 0-12 hours postoperatively (n = 8) were excluded from 12-hour analyses.

O1786 - RISK ADJUSTED VARIATION IN CICU PERFORMANCE AFTER PEDIATRIC CARDIAC SURGERY AN ANALYSIS OF THE PEDIATRIC CARDIAC CRITICAL CARE CONSORTIUM

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Background: Risk adjustment for patients undergoing congenital heart surgery is vital for comparative analyses, data transparency and public reporting. Current surgical mortality risk models focus on assessment at the time of surgery. We aimed to develop a mortality risk adjustment model to measure quality of cardiac intensive care unit (CICU) care after cardiovascular surgery, and to describe variation in performance across hospitals within the Pediatric Cardiac Critical Care Consortium (PC4).

Methods: We identified preoperative, intraoperative and variables from the first two CICU postoperative hours, which were felt to be independent of CICU management.

Variables associated with CICU mortality at $p < 0.1$ were included in a multivariable logistic regression with variables retained at $p < 0.05$. Bootstrap resampling (1000 samples) was used to derive bias-corrected confidence 95% intervals (CI) and model validation. A standardized-mortality-ratio (SMR; observed-to-expected mortality) was calculated for each hospital, and bootstrap resampling was performed again to derive the CI around the SMR estimate.

Results: The analytic cohort included 8,543 CICU admissions from 23 PC4 centers. Table 1 shows the results of the PC4 Surgical Risk Model. The model demonstrated good discrimination (C-statistic 0.92) and calibration (Hosmer Lemeshow X2 7.49, $p = 0.48$). Figure 1 demonstrates the SMR derived from the PC4 Surgical Mortality Risk Model organized by hospital CICU volume. There did not appear to be an association between hospital CICU volume and SMR.

Conclusions: The PC4 Surgical Mortality Risk Model is a novel risk-adjustment method that incorporates severity of illness variables inclusive of when care is transferred to the CICU following surgery; thus facilitating comparative analyses of CICU performance. For the first time, we report that variation in CICU performance exists across PC4 participating hospitals, with no apparent association between CICU volume and case mix adjusted survival.

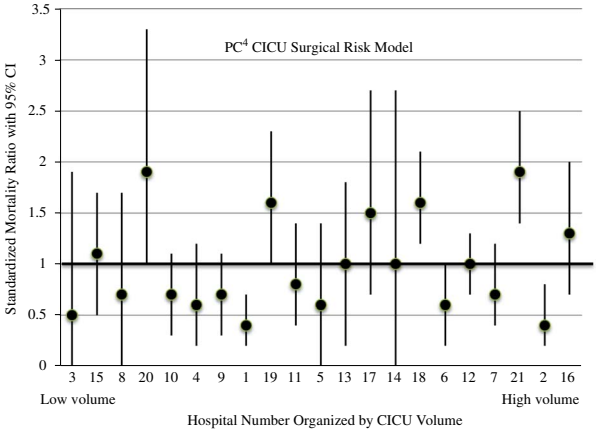

Figure.

Table.

PC[4] Surgical Risk Model

Risk Factor	Odds Ratio	p-value	95% bias corrected confidence interval
Age at surgery			
Reference: child			
Preterm neonate	4.62	<0.001	2.2 to 9.8
Term neonate	2.48	0.004	1.3 to 4.6
Infant	1.63	0.086	0.9 to 2.8
Adult	1.78	0.432	0.6 to 3.8
Any chromosomal abnormality or syndrome	1.58	0.013	1.1 to 2.3
Greater than two previous cardiac surgeries	3.05	<0.001	1.7 to 5.5
Any STS preoperative risk factor	2.13	<0.001	1.5 to 3.0
Preoperative ventilation (yes/no)	2.49	<0.001	1.8 to 3.5
STAT score	1.51	<0.001	1.3 to 1.8
On mechanical ventilation at postoperative hour two in the CICU	4.57	0.004	1.6 to 13
Maximum vasoactive index score during first two postoperative hours in CICU	1.02	<0.001	1.01 to 1.03
On ECMO during first hour following surgery in the CICU	15.88	<0.001	9.8 to 25.8

O2333 - EXTUBATION FAILURE IN INFANTS UNDERGOING PEDIATRIC CARDIAC SURGERY RISK FACTORS AND IMPACT ON OUTCOME

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Background: Extubation failure contributes significantly to morbidity costs of care following pediatric cardiac surgery.
Objectives: To identify the risk factors for failure of planned extubation after pediatric cardiac surgery and examine its impact on surgical outcomes.
Methods: Consecutive pediatric (0-18 years) cardiac surgical cases (January 2014 - March 2016) were identified from the institutional database. Failed extubation was defined as reintubation within 72 hours of extubation. A multivariable logistic regression analysis was done to identify preoperative, operative and postoperative risk factors.
Results: Of 1280 children included, 120 (9.4%) had extubation failures. 11%(138) were <30days and 46%(569) were 30days-1year old. Primary causes were cardiovascular (65, 42%) which included systemic ventricular dysfunction(27), systemic valve regurgitation (7), pulmonary hypertension(11), residual lesions(10), tachyarrhythmias(5), and pulmonary edema(5). Respiratory causes (67, 43%) were extra thoracic upper airway obstruction(18), small airway obstruction(11), lung collapse(31), and ventilator associated pneumonia(7). Other causes (23, 14%) were sepsis(10), seizures(4), apnea(18), diaphragm palsy(2), pneumothorax(2), pleural effusion (1) and re-exploration(1). Preoperative variables associated with failed extubation were age <1year, emaciated appearance, < 5th percentile of body mass index for age, oxygen saturation <85%, prematurity, and associated chromosomal anomaly. Postoperative variables were low output state and sepsis. Pulmonary artery banding (27%), Ebstein's anomaly (40%), ALCAPA (22%) and arterial switch with VSD closure and aortic arch repair (33%) had the highest incidence of failed extubation. Length of ICU stay [373 vs. 116 hours, $p < 0.001$] and mortality was 6.7% vs. 2.4%, (p-0.015) was significantly higher in patients with failed extubation.
Conclusion: Failure of planned extubation of infants is not uncommon in pediatric cardiac surgery and is associated with higher mortality and length of ICU stay. The risk is higher in younger, malnourished, premature, hypoxic infants undergoing complex cardiac surgeries.

O2571 - IMPACT OF MALNUTRITION ON THE POSTOPERATIVE OUTCOME FOLLOWING PAEDIATRIC CARDIAC SURGERY

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Background: Malnutrition is a common cause of morbidity and mortality in children with CHD. Hypermetabolic state in the peri-operative period may increase the morbidity and mortality. We analysed the impact of malnutrition in 100 children who underwent heart surgery using cardiopulmonary bypass (CPB).
Materials: In a retrospective study, 100 malnourished children, who underwent open heart surgery with CPB included. 292 children were operated between January to October 2016. Patients were divided into two groups: Simple surgery (n = 32) and Complex surgery group (n = 68). Simple group included: ASD and VSD. Complex group included: TOF, dTGA, TAPVC, Truncus arteriosus, BDG shunt, and AV canal defect. Patients were divided into Grade I to IV based on the IAP classification. Simple group: Grade I- 13, Grade II- 09, Grade III-08, and Grade IV-08. Complex group: Grade I-19 , Grade II -18 , Grade III-23 and Grade IV-02. The outcome variables measured were: Duration of CPB time, Aortic cross clamp time, duration of ventilation, length of ICU, hospital stay and in-hospital mortality.
Results: Prevalence of malnutrition in our patient population was 34.24%.The age of the patients was ranging from one month to 5 years. In Simple group 37 children had mild to moderate malnutrition (Grade I&II) and 25 children were severely malnourished (Grade III & IV). In complex group 38 children had mild to moderate malnutrion while 24 children were severely malnourished. In both groups, children with severe malnutrition required longer duration of mechanical ventilation (>72 hours) compared to children with lesser grades of malnutrition (Simple group – 9% Vs 33%, Complex group – 10.8% Vs 28%).The length of ICU and hospital stay was higher in severely malnourished children.
Conclusion: The degree of malnourishment was associated with adverse outcome following pediatric cardiac surgery. Severely malnourished children had longer duration of ventilation, ICU and hospital stay.

O2694 - USE OF NEAR INFRARED SPECTROSCOPY TO PREDICT EXTUBATION OUTCOMES IN NEONATES AFTER CARDIAC SURGERY

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Background/Hypothesis: Near infrared spectroscopy (NIRS) is used at many centers to monitor tissue perfusion after neonatal cardiac surgery. We hypothesized that an increase in NIRS values would predict extubation success in a multi-center cohort of neonates following cardiac surgery.
Methods: We conducted a secondary analysis of data collected prospectively in a study of neonates (≤ 30 days) who underwent cardiac surgery at seven tertiary care institutions in the United States in 2015. Baseline (i.e., in the operating room before surgery) and pre-extubation cerebral or renal NIRS values were analyzed. Extubation failure was defined as need for reintubation within 72 hours. NIRS measurements in neonates who extubated successfully were compared to neonates who failed extubation using student t-tests, as well as multivariate logistic regression analysis.
Results: We reviewed 283 neonates, of whom 159 had baseline and pre-extubation NIRS values recorded: 133 had cerebral and renal values, 16 had only cerebral values, and 10 had only renal values. Median age at time of surgery was 6 days (range: 1-29 days); 52 (33%) neonates had single ventricle anatomy. Fifteen (9.4%) neonates failed extubation. Baseline cerebral and renal NIRS values were not statistically different between successfully extubated neonates and neonates who failed extubation. Pre-extubation cerebral and renal NIRS were significantly higher in neonates who extubated successfully compared to those who failed extubation (Table). At the time of extubation, an increase in cerebral NIRS ≥ 5% from baseline had a positive predictive value of 98.6% (95% CI: 91.1%-99.8%) for extubation success. On multivariable analysis, an increase in cerebral NIRS by ≥ 5% was independently associated with extubation success (odds ratio 8.9, 95% CI: 1.1-73.2).

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Conclusion: Increase in NIRS values predicts extubation success in neonates following cardiac surgery. NIRS may be a helpful, non-invasive adjunct to extubation readiness assessment. Further study is warranted.

Table. Cerebral and Renal Near Infrared Spectroscopy (NIRS) Measurements.

Cerebral NIRS	Extubation Success (n = 136)	Extubation Failure (n = 13)	*p*-value
Baseline	66 ± 14	67 ± 15	0.72
Pre-extubation	70 ± 13	62 ± 12	**0.04**
Δ NIRS†	4 ± 15	-5 ± 14	**0.04**
Renal NIRS	**Extubation Success (n = 128)**	**Extubation Failure (n = 15)**	***p*-value**
Baseline	63 ± 14	63 ± 14	0.98
Pre-extubation	75 ± 14	66 ± 13	**0.02**
Δ NIRS†	12 ± 18	3 ± 20	0.09

†Δ NIRS: Baseline minus pre-extubation NIRS.

O3008 - RISK FACTORS FOR SEIZURES IN NEONATES FOLLOWING SURGERY WITH CARDIOPULMONARY BYPASS

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Objective: Neonates undergoing surgery with cardiopulmonary bypass (CPB) are at risk for seizures. Following implementation of routine postoperative continuous electroencephalography (CEEG), we reported an 8% seizure incidence of which 85% were exclusively subclinical. Determination of risk factors for post-operative seizures will help target CEEG resources.
Methods: We performed a retrospective chart review study of neonates who underwent surgery from June 2012 to December 2015 to identify electrographic seizure predictors using logistic regression. Receiving operator curve (ROC) characteristics assessed model performance.
Results: 386 of 398 (97%) of postoperative neonates were monitored with CEEG. 31 (8%) had postoperative seizures. Seizures started at a median of 29 hours (IQR 29,37) after return from the operating room (OR). Seizures were EEG-only in 28 (90%), and constituted status epilepticus in 13 (42%). Mortality was higher with (8/31, 26%) than without (22/335, 6%) seizures ($p < 0.0001$). Univariable analyses (Table) identified seizure predictors: gestational age, delayed sternal closure, deep hypothermic circulatory arrest duration, CPB duration, ECMO in the OR and cardiac arrest in the OR. On multivariable analysis (Table), gestational age <38 weeks and CPB duration >60 minutes were significant risk factors for postoperative seizures. The area under the ROC curve for this model was 0.74. If a cut-point determined by optimal model sensitivity and specificity then 291 of 386 (75%) would have been selected to undergo CEEG monitoring, and 3 of 31 (10%) with seizures would not have been identified (sensitivity 90%, specificity 26%). The 8% seizure incidence would yield a positive predictive value of 10% and negative predictive value of 97%.
Conclusion: Postoperative seizures occurred in 8% of neonates following surgery with CPB. Risk factors for seizures were gestational age <38 weeks and CPB duration >60 minutes. However, since the current model only has fair performance, development of a more robust seizure prediction model is warranted.

Table. Eleclrographic seizure predictors.

	Univariate Analysis			Multivriate Analysis	
Variable	**No Seizures**	**Seizures**	**p-value**	**Odds Ratio (95% CI)**	**p-value**
Gender					
Male	195 (91%)	19 (9%)	0.5		
Female	160 (93%)	12 (7%)			
Gestational Age (weeks)*	39 (38,39)	38 (37,39)	**0.05**	**0.4 (0.18-0.89)**	**0.03**
Identified Genetic Abnormality					
None	289 (91)	27 (9)	0.4		
Present	66 (94)	4 (6)			
Age at Surgery (days)	5 (3,7)	4 (3,7)	0.6		
Cardiac Defect					
2 V, no arch obstruction	158 (93)	11(7)	0.46		
2 V, arch obstruction	75 (94)	5(6)			
1 V, no arch obstruction	28 (88)	4(12)			
1 V, arch obstruction	94 (90)	11 (10)			
Delayed Sternal Closure					
No	291 (94)	20 (6)	**0.02**	1.16(0.43-3.16)	0.75
Yes	64 (85)	11 (15)			
Duration of DHCA (minutes)	40 (30,48)	45 (37.50)	**0.07**	1.01 (0.99-1.04)	0.07
Duration of CPB (minutes)**	46 (38,63)	61 (40.89)	**0.02**	**2.70 (1.15-6.31)**	**0.02**
ECMO in OR***					
No	349 (93%)	27 (7%)	**<0.0001**	3.52 (0.64-19.49)	0.15
Yes	6 (60%)	4 (40%)			
Cardiac arrest in OR***					
No	354 (92%)	30 (8%)	**0.03**	3.58 (0.14-90.37)	0.45
Yes	1 (50%)	1 (50%)			

Number (%) and median (interquartile range) are reported as appropriate.
CPB, cardiopulmonary bypass; DHCA, deep hypothermic circulatory arrest; OR, operating room;
CI, confidence interval; ECMO, extracorporeal membrane oxygenation; V, ventricle
*A gestational age cutoff of 38 weeks was used in the multivariate analysis.
**Cardiopulmonary bypass duration >60 minutes was used in the multivariate analysis.
***ECMO and cardiac arrest in the OR were used as these events occurred before monitoring was initiated.

INTERVENTIONAL CARDIOLOGY

O1051 - LONG TERM RESULTS AFTER IMPLANTATION OF AN UNRESTRICTIVE BABYSTENT IN THE INFANT POPULATION

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Rationale: All conventional stents suited for the implantation in newborns or infants have a maximum dilatable diameter below the expected growth of nearly all vessels in those patients. Either their use has to be avoided or later surgical removal is implicated.

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Method: The Babystent (BS) is a premounted, balloon expandable metal stent of 6 mm diameter and 20 mm length. Due to its special design the stent struts will open up along the full length of the stent from a diameter of 10 mm on, so that there is no limiting upper diameter.
Population: Between 2010 and 2014, seventeen BS were implanted in 14 patients (mean age 4 months [1–14 months]. All but one stent were placed into the aortic isthmus for coarctation. One stent was used to enlarge the superior vena cava in a 6 months old after Glenn anastomosis. Redilations were performed in all patients between one and 7 times. One BS was falsely explanted by a surgeon not realising the nature of the BS. Summarized follow-up time is 47.2 patient years ranging from 2 to 6 years.
Results: Sixteen BS are still in place in 13 patients. Circumferential integrity is lost in 5 patients with redilations of minimal 10 mm. Mean BS diameter is 8.8 mm [7–12 mm]. None of the patients has received a different stent in the position where the BS was implanted, no related re-operation was necessary. Instent tissue proliferation was found early in 5 BS, but was considered mild. A small dissection occurred in one patient after redilation.
Conclusion: The BS shows good results in treating coarctations. Despite some intimal proliferation occurred, late proliferation was rare. The loss of the circumferential integrity of the BS does not necessarily lead to collapse of the stented area.

O1081 - SAFETY AND EFFICACY OF TRANSCATHETER CLOSURE OF OUTLET TYPE VENTRICULAR SEPTAL DEFECTS IN CHILDREN AND ADULTS WITH AMPLATZER DUCT OCCLUDERS

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Background: The literature regarding transcatheter closure of outlet-type VSDs is scant.
Methods and Results: A retrospective chart review of patients with outlet-type VSD scheduled for transcatheter closure during October 2013 to November 2016 was performed at a children's hospital. Although 25 patients underwent catheterization, the intervention was abandoned in 3 because of moderate-to-severe aortic valve prolapse with aortic regurgitation (AR). Transcatheter closure was attempted in 22 patients and was successful in 20 (90.9%; 12 males and 8 females; mean [± standard deviation] age and body weight: 16.4 (± 16.0) years and 39.2 (± 24.3) kg, respectively). The median VSD diameter was 4.1 mm (range: 1.2–5.5 mm). ADO II (size range: range: 4–6 mm) was placed in 19 patients and ADO I (size: 8 mm) in only 1 patient. Device closure failed because the sheath could not be advanced through a retrograde or antegrade route in 1 patient and because the occluder dislodged in another patient. During a mean follow-up of 11.1 months (range: 1.4–25.7 months), 2 patients showed trivial-to-mild new-onset AR on echocardiography. Preprocedural mild AR disappeared in 4 patients. No heart block, or device failure occurred.
Conclusions: Transcatheter closure of outlet-type VSDs with Amplatzer duct occluders is feasible. Although no significant aggravation of AR was observed in the mid-term follow-up, long-term follow-up is mandatory.

O1123 - RIGHT VENTRICULAR OUTFLOW TRACT STENTING IN TETRALOGY OF FALLOT INFANTS WITH RISK FACTORS FOR EARLY PRIMARY REPAIR

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Background: Tetralogy of Fallot (TOF) with cyanosis requiring surgical repair in early infancy, reflects poor anatomy, and is associated with more clinical instability and longer hospitalization than those who can be electively repaired later. We bridged symptomatic infants, with risk factors for early primary repair, by right ventricular outflow tract (RVOT)-stenting (Stent).
Methods and Results: Four groups of TOF with confluent central pulmonary arteries were studied: Stent (n = 42), primary repair (<3 months-old) with pulmonary stenosis (early-PS; n = 44) and atresia (early-PA; n = 49) and Surg > 3mo (3-11 months old) (n = 45). Stent patients had the smallest pulmonary arteries with a median [95% credible intervals] Nakata index (mm2 /m2) of 79 [66, 85] compared with the early-PA 139 [129, 154], early-PS 136 [121,153] and Surg > 3mo 167 [153, 200] groups. Only Stent infants required unifocalization of aortopulmonary collaterals (17%). Stent and early-PA infants were younger age and lower weight than earlyPS infants. Stent infants had the most multiple comorbidities. Stenting allowed deferral of complete surgical repair to an age (6 months), weight (6.3 [5.8, 7.0] kg) and Nakata index (147 [132, 165]) similar to the low-risk Surg > 3mo group. The 3 early treatment groups had similar ICU/hospital stays and high re-intervention rates in the first 12 months after repair, compared with the Surg > 3mo group.
Conclusions: RVOT-stenting of symptomatic TOF with poor anatomy (small pulmonary arteries) and adverse factors (multiple comorbidities, low weight) relieves cyanosis and defers surgical repair. This allowed pulmonary arterial and somatic growth with clinical results comparable to early surgical repair in more favourable patients.

O1411 - IMPLANTATION OF A NOVEL ATRIAL FLOW REGULATOR DEVICE IN SEVERE PULMONARY ARTERIAL HYPERTENSION

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Background: Balloon atrial septostomy and atrial septal stenting are accepted modalities to enhance systemic cardiac output in severe pulmonary arterial hypertension (PAH) though the size of the atrial septal communication is unpredictable. An atrial flow regulator (AFR) device has been designed with varying sizes of fenestrations with the aim to provide a predictable atrial communication.
Materials andMethods: Patients with syncope and or right heart failure due to severe Idiopathic PAH were planned for AFR device implantation with 8 or 10 mm fenestration after transseptal puncture, as part of an investigational trial.
Results: 9 patients (two males) with severe Idiopathic PAH underwent AFR device implantation during June to December 2016. Age ranged from 15 – 39 years (median 30). All patients had history of syncope . All except one was in NYHA Class 3. All were receiving optimal pulmonary vasodilator therapy and were non-responders on vasodilatory testing. Fenestration size was 10 mm in

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six patients and 8 mm in three. There were no procedural complications. Right to left shunt was demonstrated soon after implantation in all. Arterial oxygen saturation postprocedure ranged from 87 to 92% (mean 90).Mean cardiac output improved from 3.47 to 4.32 L/mt.All patients were maintained on antiplatelet agents. At follow up ranging from 2 days to 6 months, none had recurrence of syncope, right to left shunt was sustained in all. Mean six minute walk distance improved from 375 m to 427 m. All except one had reduction in the N terminal Pro BNP levels.
Conclusion: Atrial flow regulator device implantation is a novel and safe method of creating a predictable interatrial right to left shunt in patients with severe PAH and syncope. Long term follow up is needed to document the patency of the fenestration.

O1417 - PERCUTANEOUS CLOSURE OF PATENT DUCTUS ARTERIOSUS WITH THE NIT OCCLUD® PDA DEVICE IN 268 CONSECUTIVE CASES

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Background: We report our single-center experience with the pfm Nit-Occlud® coils for closure of patent ductus arteriosus (PDA).
Methods: From 08/2008 to 12/2015, 268 consecutive patients were admitted for transcatheter closure of a PDA with the pfm Nit-Occlud® coil. Clinical, echocardiographic and angiographic data were evaluated retrospectively.
Results: Median age at the time of the intervention was 5.2 years (range 5 month to 62 years and median weight was 19.3 kg (range 5.5 to 97 kg). 10 patients (3.7%) had a body weight <10 kg. The most common PDA types were type E and A, according to the Krichenko classification (44.4% and 33.2%). 12 cases were residual PDAs postoperatively (4.5%). Mean diameter at the narrowest point was 1.8 ± 0.8 mm (range 0.4 to 4 mm), mean size of the ampulla was 5.2 ± 2.7 mm (range 1 to 15 mm) and mean length was 9.2 ± 3.3 mm (range 2 to 25 mm). Mean pulmonary artery pressure before closure was 30.7 ± 4.4 mmHg (range 20 mmHg to 47 mmHg). Device implantation could be successfully achieved in all cases. Device sizes applied were 4 × 4 mm (17 pt) and up to 11 × 6 mm (21 pt). The 7 × 6 mm device was most commonly used (37.6%). Mean fluoroscopy time was 9.3 ± 7.3 min; mean stay in hospital was 3.4 ± 1.5 days. In one patient a second coil had to be placed. In a further patient with headache post-interventionally, MRI verified a minor ischemic stroke. Symptoms resolved completely during follow-up with 7 days of low-dose aspirin. No further adverse events occurred. Closure rates documented at 3-10 days, 3 month and 6 month post intervention were 95.1%. 97.6 and 98.4%, respectively.
Conclusion: Closure of PDAs with sizes up to 4x15 × 9 mm (min. diameter/ampulla/length) can be done effectively and safe using the Nit-Occlud® PDA device. 6-month follow-up showed an uneventful outcome and excellent closure rates.

O1433 - TRANSCATHETER CLOSURE OF PATENT DUCTUS ARTERIOSUS UNDER ECHOCARDIOGRAPHIC GUIDANCE

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Background: Transcatheter closure of patent ductus arteriosus (PDA) is done in the cardiac catheterization laboratory, usually under fluoroscopic and angiographic guidance. The aortogram is used for assessing PDA size and shape pre-device implantation. Additionally, post-device deployment aortograms are applied for the assessment of device position, profile and residual shunt. Angiograms expose patient to radiation and possible untoward effects of contrast media. Recently, transthoracic echocardiography {TTE} has been utilized to guide in PDA closure to avoid radiation exposure, reduce the risk of arterial sheath insertion, as well as contrast material.
Material and Method: Forty six patients {30 Females and 16 Males} underwent TTE guided device closure of PDA between July 2013 and November 2016. Age range 4 months to 8 years and weighing 3.2-23 Kgs . In all the patients the procedure was performed under conscious sedation except one was intubated and ventilated. In 37 patients the procedure was performed transvenously and in 10 retrogradely.
Results: The device was successfuly deployed in all patients except in one the device was embolized to pulmonary artery but successfully retrieved and replaced by a bigger one. Flouroscopy time ranged between 2.2 and 13 minutes.
Conclusion: Transcatheter closure of PDA can safely and effectively be performed under TTE guidance, it can be completed transvenously or retrogradely , we prefer the transvenous approach so that we do not need to pass a sheath through the femoral artery.

O1649 - LONG TERM OUTCOME FOLLOWING PERCUTANEOUS CLOSURE OF ISOLATED SECUNDUM ATRIAL SEPTAL DEFECTS IN CHILDREN A FRENCH NATIONWIDE SERIES OF 1000 CONSECUTIVE PATIENTS

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Introduction: Transcatheter closure has become the preferred treatment strategy in most cases of isolated, secundum atrial septal defect (ASD). Although widely used, data on long-term outcomes in the pediatric population are scarce. We aimed to assess procedural characteristics, early clinical outcome, long-term device-related complications and the electrical remodeling after transcatheter closure of isolated ASD in children.
Methods: A 1998-2014 retrospective multicentre study was performed at 8 French tertiary institutions, including all patients <18 yo who attempted a percutaneous ASD closure with an Amplatzer Septal Occluder.
Results: 1000 children [38% males, median age: 9 yrs (0.7-18.0), median weight: 27 kg (6-92)] were referred for transcatheter ASD closure. They all had a significant left-to-right shunting assessed by right ventricular dilation and/or a 1.5:1 Qp/Qs ratio; Median

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ASD size was 15 mm in transthoracic echography (TTE). ASD closure was guided by fluoroscopy and transoesophageal echocardiography in 627 cases (62.7%) or TTE in 373 cases (37.3%). Procedural success rate was 94% with a median occluder size of 19-mm (4-40). Device placement was unsuccessful in 60 patients (6%) due to unfavourable anatomy in 38, early device embolization in 12 and other causes in 10 patients. Follow-up data were available for 829 patients. After a mean FU of 53 ± 31 months (range, 5–204), all patients were alive and 96% were asymptomatic. Long-term complications included supraventricular arrhythmias (n = 6) and pulmonary hypertension (n = 2). No cardiac erosion, late ASO dislodgement or stroke occurred.From an electrical standpoint, supraventricular arrhythmias occurred in 6 patients, no late atrioventricular block was observed and there was no significant difference between preprocedural and last follow-up ECG.
Conclusions: Our large-scale pediatric cohort confirms that transcatheter closure of isolated secundum ASDs is a safe procedure in children, with a favourable long-term outcome and no life-threatening delayed complication.

O1759 - FEASIBILITY OF TRANSCATHETER PULMONARY ARTERY BANDING AND LONG TERM OUTCOMES IN COMPARISON TO SURGICAL BANDING IN A SWINE MODEL

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Background: There have been attempts to develop a transcatheter pulmonary flow restrictor (PFR) as an alternative to surgical pulmonary artery banding (PAB) without great success. We modified the Microvascular-Plug (MVP) to be used as a PFR. The objectives of this study were to demonstrate feasibility of transcatheter implantation and retrieval of this PFR, and compare long term PA growth whilst using the PFR vs. PAB.
Methods: The PFR was implanted in bilateral PA in 8-newborn piglets weighing 2.12-3.17 kg. A left PAB was placed via thoracotomy in 4 other newborn piglets (2.29-2.84 kg). Transcatheter retrieval of the PFR was performed after 3 weeks (4.6-5.6 kg) in 3-piglets, 6 weeks (9.4-10.6 kg) in 3-piglets and at 12 weeks (19.2-20.4 kg) in 2-piglets. Four piglets were sacrificed following PFR retrieval for histopathology. The rest were followed until 12 weeks to study PA growth. Debanding of PAB was performed at 6 weeks (8.1-11.4 kg). The surgical control piglets were recatheterized at 12 weeks (23.4-26 kg) to compare PA growth.
Results: The median time required for PFR implantation and retrieval were 40 and 90 minutes respectively. Immediately post-PFR implantation, the RV systolic pressure increased from a median of 20 mmHg to 51 mmHg (40-67 mmHg; $p < 0.001$). There was 75% endothelialization of the device by 6weeks. Retrieval of PFR was 100% successful at 3weeks, 75% at 6weeks and 50% at 12weeks. At 12 weeks, the LPA size in the PFR group and the PAB group were similar (median 8.6 vs. 8.4 mm, $p = 0.44$). There was grade-1 vessel injury immediately post-retrieval and none at 12weeks.
Conclusions: It was feasible to use the MVP as a PFR. Transcatheter retrieval of this device is possible before complete endothelialization. PA growth was not affected post-retrieval and is comparable to surgical PAB. This PFR has to be trialed in humans before recommending its routine use.

O1840 - STENT PLACEMENT FOR TREATMENT OF AORTIC COARCTATION IN CHILDREN UNDER 30 KG ACUTE AND LONG TERM OUTCOMES

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Introduction: The use of stents for aortic coarctation (CoAo) in children presents some challenges as limitations of vascular access and lack of published data regarding the reintervention. We sought to evaluate the safety, feasibility and effectiveness of these stents in children, with an emphasis on follow-up.
Methods: A retrospective analysis of children under 30 kg who had undergone stent placement for CoA between April/2009 and December/2015 was performed. Stents that can be expanded to larger diameters have been used. Demographic, clinical, hemodynamic and follow-up data were collected. The endpoints evaluated included: severe adverse events (SAE), persistent high blood pressure and need for reintervention.
Results: Thirty-seven patients (27 with native CoAo), with mean age and weight of 5.4 ± 3.4 years and 16.2 ± 7.0 kg were enrolled respectively. Immediately, the peak-peak gradient decreased from 33.7 ± 15.1 to 5.4 ± 5.3 ($p < 0.01$) and the ratio of CoA diameter/Descendent Aorta increased from 0.40 ± 0.16 to 1.0 ± 0.20 ($p < 0.01$). There were no deaths or immediate SAE. Thirty-four patients were followed during a mean period of 43.1 ± 19.4 months, using echocardiography and computed tomography of aorta. Five patients still needed antihypertensive drugs. Seven patients required percutaneous reinterventions (36.1 ± 19.0 months after initial treatment) due to aortic aneurysm (1), residual stenosis above the stent (1) and adjustment to somatic growth (5). One patient required surgery due to residual hypoplasia of aortic arch (15.1 months later). Freedom of all reinterventions was 95%, 78%, 54% and freedom of balloon dilations was 95%, 81%, 81% in 12, 36 and 60 months, respectively. All of reinterventions have been carried out successfully, with no SAE.
Conclusion: Stenting for treatment of CoAo in children was feasible, safe and effective. A significant rate of reintervention was observed mainly due to the anticipated need for stent diameter adjustment to the somatic growth. However, continuous surveillance for arch hypoplasia and aneurysm formation is advisable.

O1852 - PROSPECTIVE STUDY ON PATENT DUCTUS ARTERIOSUS STENTING IN CYANOTIC CONGENITAL HEART DISEASE WITH DUCTUS RELATED BRANCH PULMONARY ARTERY STENOSIS

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Ductus-related branch pulmonary artery (PA) stenosis is common in cyanotic congenital heart disease (CHD). Ductal stenting is not recommended for concerns of jeopardizing growth of the affected branch. This however has not been based on extensive evidence.
Objective: To study PA growth in duct-dependent cyanotic CHD with branch PA stenosis following ductal stenting.
Methods: Prospective, non-randomized study. Inclusion criteria: Patients with duct-dependent pulmonary circulation age

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<3 month. Pre-procedure CT Angiogram (CTA) was performed for morphologic evaluation of PDA and PA. Ductal stenting was performed in all patients except in those with severely tortuous PDA. PA growth was monitored by CTA at 3 months and cardiac catheterization at 9 months. Affected branch PA (P1) and contralateral PA (P2) were measured. P1/P2 ratio was calculated.
Results: Between February 2014 to May 2015, 34 patients underwent PDA stenting, 16 via transfemoral artery approach (47.1%), axillary artery in 12(35.3%), transvenous in 5(14.7%). 26 (75.5%) had LPA stenosis, 5(14.7%) RPA stenosis. 28 had CTA evaluation at 3 months and 25 had catheterization at 9 months. At presentation, P1 was 3.5 + 3.0mm (Z score -1.48) and P2 4.6 + 3.6mm (Z score -0.31) respectively, P1/P2 ratio 0.7. At 9 months, P1 was 7.3 mm (Z score +1.68) and P2 9.7 mm (Z score +2.84), P1/P2 ratio 0.8. 3 patients required rescue Blalock-Taussig shunt due to compressed stents and 1 stent thrombosis. 2 stents redilated for in-stent stenosis. There were no deaths and no stent migration. 13 underwent corrective surgeries and 6 underwent palliative cavopulmonary shunt.
Conclusions: Ductal stenting in ductus-related branch PA stenosis results in satisfactory growth of PA despite jailing of an affected branch. This is a viable alternative to surgery for the early palliation of duct-dependent cyanotic CHD. However close monitoring is required and further studies are warranted.

O1907 - EVALUATION OF CENTRAL PULMONARY ARTERIES GROWTH FOLLOWING PDA STENTING IN PATIENTS WITH DUCT DEPENDENT PULMONARY CIRCULATION

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Background: If stent is implanted in arterial duct,it may result in better angle between PDA and pulmonary arteries, allowing for improved growth.
Aim of Work: evaluate ability of PDA stenting to promote pulmonary arteries growth in patients with congenital heart diseases-duct dependent pulmonary (CHD-DDP) circulation.
Methods: 58 patients with CHD-DDP who had PDA stenting 2009-2016 in Mansoura and Tanta University children Hospitals underwent evaluation of pulmonary arteries (within 6 -12 months post stenting).
Results: PDA stenting was successful 49/58 (84.5%) patients; 31 males (63.3%) 18 females (36.7%) mean age 13.5 ± 10.4 days, mean weight 2.9 ± 0.5 kg.22 (44.9%) patients had complex CHD-DDP, 14 (28.6%) PA/IVS, 13 (24.5%) PA-VSD. Prestenting, presecond stage measures were compared.oxygen saturation increased 67.1 ± 8.0 to 81.2 ± 8.0 (P <0.001*). RPA increased 0.36 ± 0.05 to 0.60 ± 0.11, P <0.001 *(Z-1.23 ± 0.91 to 0.18 ± 0.81 (P <0.001*) LPA increased 0.34 ± 0.06 to 0.58 ± 0.10, P <0.001* (Z-1.13 ± 0.86 to 0.48 ± 0.97, P <0.001*). Nakata index increased 108.89 ± 33.53 to 183 ± 48 ± 40.58 mm^2/m^2 (P < 0.001*)
Conclusion: PDA stenting is effective in promoting global and individual pulmonary artery growth in CHD with duct dependent pulmonary circulation. It ensures uniform balanced distribution and growth of pulmonary arteries.

O2076 - PERCUTANEOUS CLOSURE OF ATRIAL SEPTAL DEFECT USING NIT–OCCLUD® ASD–R LONGTERM RESULTS

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Background: Ancient weaving technique inspired the development of closure devices for congenital heart diseases.
Aim: To assess clinical efficacy and safety of Nit–Occlud® ASD–R in the percutaneous closure of secundum atrial septal defects (ASD). Background: ASD is 2.3 times more frequent at high altitude. Percutaneous ASD closure has been demonstrated to be safe and effective with many potential benefits over surgical closure.
Methods and Results: Retrospective, observational study of patients treated with the Nit–Occlud® ASD–R between May 2007 and March 2011. Fortynine out of 53 patients underwent successful device closure of ASD. The median age at closure was 11 years (range 3–67) and the mean weight was 27,1 kg (range 13–75). Sixtynine per cent of patients were females. The mean fluoroscopy time was 14 minutes (range 5–53). The mean procedure time was 70 minutes (range 45–150). The mean defect size measured by the stop –flow technique was 17,8 mm (range 5.6–31) with, the mean size of the implanted device being 18 mm (range 6–28), 1.0 times the defect size. Patient follow up up to Dezember 2017 ranged from 67 to 113 months, median 87 months. Immediate closure was observed in 71.4% of the cases, with 91.7% closure after 24 hours, 93.7% after 1 month, 95.8% after 3 months and 100% after 6 months. Device embolization occurred in one patient within 24 hours requiring surgical device removal. There were no other major complications including death in the follow up time.
Conclusions: Our experience with the NitOcclud® ASD–R device demonstrates it to be safe and highly effective with very.

Figure 1.

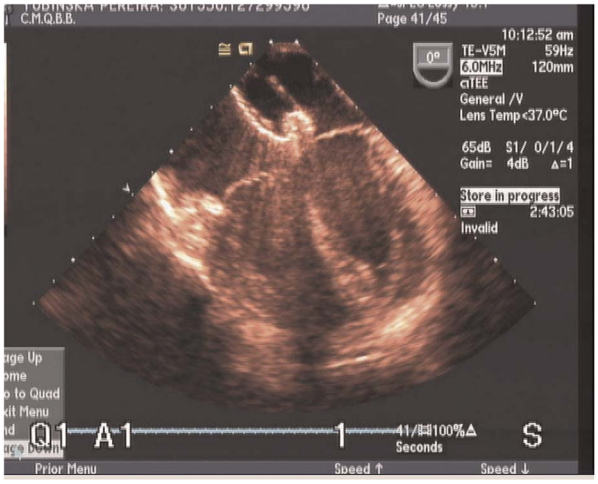

Figure 2.

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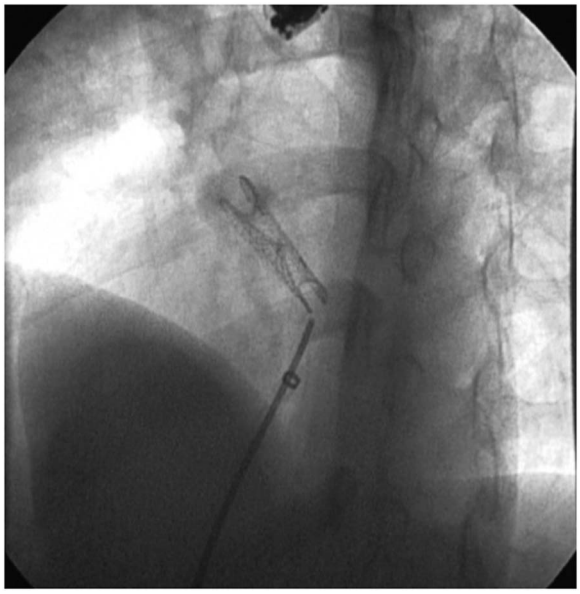

Figure 3.

O2110 - TRANSCATHETER CLOSURE OF THE PATENT DUCTUS ARTERIOSUS IN PREMATURE INFANTS IS AN EFFECTIVE ALTERNATIVE TO VIDEO ASSISTED THORASCOPIC SURGICAL CLOSURE

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Background: Patent ductus arteriosus (PDA) closure in premature infants has been primarily surgical. Transcatheter closure provides a less invasive alternative. Video-Assisted Thorascopic Surgery (VATS) and transcatheter PDA closure in premature infants are compared.
Methods: Medical records of premature infants under 3 kilograms, with VATS or transcatheter PDA closure from 1/2012 to 1/2017, were reviewed. Procedural details, complications, and short-term outcomes were compared.
Results: Patient data and results are presented in Table 1. Thirty-five infants underwent VATS; Twenty infants underwent transcatheter closure with a Medtronic Microvascular Plug (n = 16) or Amplatzer Vascular Plug II (n = 4), deployed prograde via a 4 F delivery system, without arterial access. Patients of each group were of similar age, but weighed less at time of surgery. Complete PDA closure was achieved in all patients except one VATS patient with a small residual shunt 70 days after surgery. Procedural complications for VATS included: 3 with vocal cord dysfunction; and 1 each with peri-procedural cardiopulmonary resuscitation, chylothorax, and post-operative hypertension. No acute procedural complications occurred in the catheterization group. One transcatheter device embolized 9 days after deployment and was successfully retrieved and another implanted. Fewer complications occurred in the catheterization group. Time to extubation and discontinuation of respiratory support was similar. Tracheostomy was performed in 2 VATS patients and 1 catheterization patient. Surgical patients were more likely to be discharged on home oxygen therapy. There were 3 deaths in the VATS group, with none attributed to the procedure.
Conclusions: Transcatheter device closure of PDAs offers a successful and less invasive alternative to surgery in premature infants with fewer complications. Extubation and freedom from respiratory support were comparable to VATS, nevertheless, fewer patients required home oxygen at discharge. Transcatheter closure warrants further study and development.

Table.

	Catheterization	Surgery	p-value
Gestational age (weeks)	27.3 (23-35.6)	25 (23.4- 36.3)	0.07
Birth weight (kg)	0.84 (0.57-2.49)	0.72 (0.42-1.88)	***0.02***
Age at procedure (days)	52 (15-108)	49 (20-94)	0.3
Weight at procedure (kg)	2.2 (1.3-2.8)	1.3 (0.71-2.01)	***<0.001***
Procedural complication	0/20	7/35	***0.04***
Time to extubation (days)	5 (0-29)	6 (0-47)	0.12
Time to discontinue O_2 (days)	14 (0-153)	24 (0-118)	0.99
Discharge on O_2 therapy	1/17	15/31	*0.003*

Data presented as median (range), except categorical data

O2111 - CHALLENGES OF DEVICE CLOSURE OF PATENT DUCTUS ARTERIOSUS WITH PULMONARY ARTERY HYPERTENSION

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Introduction: Transcatheter Device closure of patent ductus arteriosus (PDA) is treatment of choice. However device closure in presence of severe pulmonary artery hypertension (PAH) is challenging. But there is limited literature on patient selection, technical considerations, and complications of device closure in patients of PDA with severe PAH.
Aim: To know the challenges and evaluate techniques for feasibility, efficacy of device closure of PDA with severe PAH.
Materials and Results: Out of 1,695 cases of device closure of PDA, 296 (17.5%) with PAH formed material for this study. To test the feasibility of device closure by occlude PDA with the appropriate device, instead of balloon for ten minutes without oxygen inhalation. The device was released only if PAH(especially mean pressure) reduced. PAH decreased in all except in 1 patient after closure with muscular ventricular septal occluder (MVSDO), pulmonary artery pressure (PAP) transiently increased (became suprasystemic), without significant reduction in aortic pressure. Device embolized in 10 patients (3.4%). Percutaneous retrieval was done in 5 (by snare in 3 and by fixing the cable to device in 2) and replaced with bigger devices. The surgical removal of the embolized MVSDO and ligation was done in 5 cases. All patients were on oral sildenafil and bosentan until PAP regressed to normal. Follow up was from 6 months to 9 years. No residual shunt in any patient on follow-up. The PAP regressed to normal in all except 5 cases (2.03%) of Down's syndrome with systemic PAP.
Conclusions: Device closure of PDA with PAH is feasible, safe in all age groups. Temporary PDA occlusion with device is effective and time saving for evaluating pulmonary vascular reactivity. Device embolization in aorta is higher with severe PAH. Novel method of retrieval is effective.

O2115 - PERCUTANEOUS TRANSCATHETER CLOSURE OF ATRIAL SEPTAL DEFECT ONLY UNDER THE GUIDANCE OF TRANSESOPHAGEAL ECHOCARDIOGRAPHY IN CHILDREN

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Objective: To investigate the feasibility, safety and effectiveness of transcatheter closure of atrial septal defect (ASD) under the guidance of transesophageal echocardiography (TEE) in children.
Methods: From Nov.2014 to Aug. 2016, 129 cases of children undergoing percutaneous transcatheter closure of atrial septal defect received the procedure completely guided and monitored by TEE. After 3 month of the operations, patients were followed up by conducting transthoracic echocardiography.
Results: 129 cases were identified by TEE as having atrial septal defect, The diameter of ASD was 3~24mm, the median was 9.52±4.12mm, the diameter of the Amplatzer occluder was 6~ 32mm, the median was 14.19±4.60 mm.Immediate post-operative check by TEE and during the follow-up by TTE, there were no complications such as occluder displacement, valvular injury, peripheral vascular injury and pericardial effusion. Residual shunt about 1.7mm was documented in a 2 holes patient and self-healed after 1 month. Residual shunt about 3 mm in another multiple holes patient existed during follow-up.
Conclusions: Percutaneous transcatheter closure of atrial septal defect completely guided by TEE is safe and effective. The advantages of high success rate, low complication rate and reasonable price make it easy to promote in children.

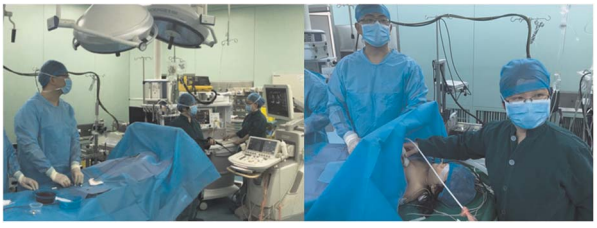

Figure 1.
Operating in the OR only under the guidance of Echo, no DSA, no X-Ray.

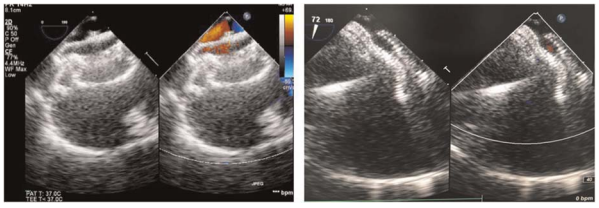

Figure 2.
The interval rim between two holes was 8.3 mm, two Amplatzer occluders were implanted, only under the guidance of Echo.

O2156 - CHEATHAM PLATINUM COVERED STENT IN AORTIC COARCTATION 13 YEARS SINGLE CENTER EXPERIENCE AND FOLLOW UP

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Background: Stent implantation is an effective therapy for aortic coarctation and recoarctation. Covered stent implantation may provide protection from potential complications. Here we report on our 13 years' experience with the use of covered Cheatham-Platinum (CCP) stents.
Methods: From January 2003 and January 2017, 122 patients with native (80 pts) or post-operative (42 pts) aortic coarctation were treated at our institution. At procedure median age was 16,5 years (range 6-68 years) and median weight was 58 Kgs (range 22-110 kg). The median gradient before the treatment was 40 mmHg (range 12-75 mmHg). Seventy-seven subjects had complex lesions (75.5%) while 25 patients had simple lesions (24.5%).
Results: Success rate was 95% (72,5% with a residual gradient <10 mmHg; 20,5% with a residual gradient between 10 and 20 mmHg). Mean coartaction gradient decreased from 39.7 to 5,7 mmHg ($p < 0.001$). Mean coarctation segment diameter increased from 5.7 to 13.4 mm ($P < 0.0001$). Incidence of early related-procedure adverse events was 8%. (lesion of external iliac artery, transient ischemia of left hand, transient femoral artery spasm, hyponatremia and convulsion). Two cases with aortic rupture after bare stent implantation or redilatation were treated. In both cases, covered stent implantation was used as a rescue option. At a median follow-up of 53 months (range 12-164 months) reintervention occurred in 21 patients (17%). Independent predictors associated with reintervention at follow up included the presence of complex lesions (P 0.0054) and ratio proximal aorta/coartaction (OR 1.06; CI: 1.01 -1.11, P 0.009). Anti-hypertensive medication was needed in 39 subjects (while it was needed in 102 subjects pre-procedure).
Conclusions: Covered stent implantation for aortic coarctation is a safe and efficacious treatment. Covered CP stents are rescue devices in case of aortic rupture.

O2351 - PERCUTANEOUS PULMONARY VALVE IMPLANTATION WITH SAPIEN VALVES IN NATIVE AND LARGE RVOT; EARLY AND MID TERM RESULTS

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Introduction: We present early and midterm results of PPVI with Sapien valve in repaired tetralogy of Fallot (TOF) patients with native-large RVOTs.
Method: 62 s/p repaired TOF patients who had native RVOT with with free pulmonary regurgitation, significant dilatation of the RV and without significant RVOT stenosis. Balloon sizing was performed with compliant (34 mm Amplatzer sizing) for interrogation (BI). The size of the balloons that the Andra Stents XXL would be mounted on was decided up to the indentation diameter occurred during BI; as at least 1 mm larger than the indentation diameter.
Results: Mean age and weight of the patients were 17.7 (7-50) years and 42(22-90) kg, respectively. Before presenting mean pressure gradient between RV and MPA was 4.8 (0-10) mmHg. Mean indentation diameter with BI was 26.4(22-32) mm. Mean balloon size used for prestenting was 28.1(24-30) mm. Successful PPVI was achieved in all; 29 mm in 53 and 26 mm in nine. PPVI was performed in same session in seven and 3-12 weeks after prestenting in 55. In addition to RVOT stenting, stent implantation to branch pulmonary artery was performed in 13. There was no significant complication. Valve function was good in all immediate after and at the last follow-up; a median of 14 months (2-29 months). RV volumes decreased and mild paravalvar leakage was observed only in six. No reintervention required yet.
Conclusion: PPVI with Sapien valve, which has larger sizes as 26 and 29 mm, is feasible and safe in patients with native RVOT

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without stenosis. Prestenting for providing a secure landing zone is the most important part of the procedure. Only Andra XXL stents which has an expansion capacity up to 32 mm can be used for this purpose, currently.

O2359 - TRANSCATHETER CLOSURE OF CORONARY ARTERY FISTULAS; SINGLE CENTER EXPERIENCE IN 9 YEARS PERIOD

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Introduction: We present our experience with CAF's between 2007 and 2016.
Method: 29 patients aged between 2 months - 67 years (median 7 years) underwent to cardiac catheterization. The procedure was performed preferably with retrograde approaches if it is feasible, otherwise antegrade approach was used by establishment an AV loop.
Results: In 29 patients, angiograms demonstrated 31 fistulas arising from right coronary artery (RCA) (13), left anterior descending artery (8), circumflex artery (CX) (5), left main coronary artery (5). They were opening to right ventricle (14), right atrium (12), pulmonary artery (3), superior vena cava (1) and bronchial artery (1) with single (19) and multiple orifices (12). There were 7 fistulas with multiple feeding arteries. Transcatheter closure was not attempted since they were very small in six. In two patients with two separate fistulae, large one was closed and the other small ones were left untreated. Intervention was successful in all and realized with retrograde and antegrade way in 20 and 3, respectively. Complete occlusion was achieved in all. 8 coils, 14 vascular plugs (1, 2 and 4) and 4 duct occluders were used. A patient suddenly died four days later probably due to thrombosis in huge and slow filling coronary arteries since the reciprocal competitive flow after closure. There were huge dilated RCA and CX artery communicating with each other acting as multiple feeding arteries and opening into the RV. It was closed by a vascular plug just distal to the orifice. Other patients are well without recanalization during the median 41 months of follow-up.
Conclusion: CAF's may present in a great variety in morphology. It is not uncommon to see multiple feeding arteries and especially multiple distal openings. Effective and safe percutaneous transcatheter closure is possible in majority of cases but it is not free of complications.

O2479 - BRANCH PULMONARY ARTERY VALVE IMPLANTATION REDUCES PULMONARY REGURGITATION AND IMPROVES RIGHT VENTRICULAR SIZE FUNCTION IN PATIENTS WITH LARGE RIGHT VENTRICULAR OUTFLOW TRACTS

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Background/Hypothesis: Patients with large right ventricular outflow tracts (RVOTs) are excluded from available percutaneous pulmonary valve options using standard techniques. In some of these patients, percutaneous branch pulmonary artery (BPA) valve implantation may be feasible. However, the long term effects of valves in the BPA position is largely unknown.
Materials and Methods: Retrospective data from 10 international centers was collected on patients with significant pulmonary regurgitation in whom a percutaneous BPA valve was attempted.
Results: 22 patients, age 26 (13-58) years, weight 72 (26-147) kg underwent an attempt at percutaneous BPA valve implantation (14 unilateral and 8 bilateral). Melody valve implants were attempted in 18 patients and Edwards Sapien valves (all unilateral) in 4 patients. There were 2 failed attempts, with both patients eventually undergoing successful percutaneous valve implants (1 in a BPA and 1 in the native RVOT). Procedural complications occurred in 2 patients (wire related lung injury in 1 and pulmonary emboli in 1). Follow up echocardiography demonstrated no patient having more than mild regurgitation through the implanted valves with 50% of patients showing a reduction in right ventricular size. MRI data (11 patients) revealed a decrease in RVEDV index from 157 (103-429) to 90 (76-429) ml/m^2 ($p < 0.01$) and a decrease in RVESV index from 80 (41–387) to 47 (41- 347) ml/m2 ($p = 0.015$). At a follow up of 21 (1-91) months, NYHA functional class improved from 2.6 (+/- 0.9) to 1.7 (+/- 0.8), ($p < 0.01$), with 4 late deaths. One valve was surgically explanted due to endocarditis.
Conclusions: Percutaneous BPA valve implantation results in a reduction in RV volume with favorable clinical benefit in the intermediate term. Until percutaneous valves for large RVOT become widely available, this procedure remains an option for select patients.

O2522 - RADIATION IN CONGENITAL CARDIAC INTERVENTIONS ELEVEN YEARS OF EXPERIENCE IN A TERTIARY CENTRE.

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Background and Aim: Exposure to ionising radiation is one of the important adverse effects of cardiac catheterisation. Children are uniquely more susceptible to the potential carcinogenic effects as their tissues may be more sensitive (as they are actively growing) and they have many more years or life following the exposure for the effect to be seen. This study sought to report patient radiation dose across a range of interventional cases in a tertiary paediatric cardiac centre.
Method: All patients who underwent an interventional procedure in our supra-regional centre between 01/01/2005 to 31/12/2015 were identified from the Departmental electronic databases. Measures of radiation exposure including fluoroscopy (screening) time (minutes), DAP (Dose-area product) in cGy.cm2 (centigray. square centimetre) basic demographic data like patient's procedural age and weight, procedural type and type of interventional procedure were recorded in a custom-designed database.

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The effective dose (in milliSievert; mSv), was calculated using PCXMC software (v2.0, STUK, Helsinki, Finland).

Results: 3165 procedures were performed. Details in Table 1–3. For overall cohort, the median fluoroscopy time was 16 minutes (IQR 5–20 minutes), Median DAP was 203 (IQR 40 – 1100) cGy.cm2. The median Effective dose was 2.24 mSv (IQR 0.8 to 6.1 mSv ICRP 103). Overall, nearly 16% of the cases had DAP of more than 2000 cGy.cm2 of skin exposure. The Kruskal-Wallis test showed that there were statistically significant differences in all measures of radiation (Screening time, DAP and effective radiation dose) exposure across procedure types ($p < 0.0001$). Para valvar leak occlusion cases were associated with the highest median effective dose, followed by fenestration closure in Fontan and VSD occlusion.

Conclusion: This study provides a guide ranges for radiation dose and fluoroscopy time for common procedures. They serve as benchmarking tools and also provide a reference standard for which future dose reduction interventions can be compared.

Table 1. Demographic and Procedural characteristics of the cohort (n = 3,165)

Median Age (range)	3.75 y (0–82yr.)
Median Weight (range)	15kg (1–147)

Procedures	Number	Percentage
Diagnostic Procedure	608	19.21%
Atrial communication device occlusion	536	16.94%
PDA Occlusion	417	13.18%
Pulmonary valvuloplasty	247	7.80%
Atrial Septostomy	219	6.92%
Coarctation Balloon/Stent/Redilatation	210	6.64%
Pulmonary Artery Angioplasty/stent	196	6.19%
PDA Balloon/Stent	121	3.82%
Aortic Valvuloplasty	103	3.25%
Extracardiac Vascular occlusion by coil/device	93	2.94%
Miscellaneous (Balloon/Stenting)	78	2.46%
Pericardiocentesis	49	1.55%
Airway Procedure	47	1.48%
Pulmonary vein angioplasty - Stent	34	1.07%
RVOT dilatation/Stent	32	1.01%
Intervention on systemic-pulmonary shunt	27	0.85%
Vascular Access	24	0.76%
Para valvar Leak Occlusion	22	0.70%
Pulmonary Valve RF Perforation	21	0.66%
VSD Closure	20	0.63%
Interventional Valve Placement	18	0.57%
Atrial Septal Stent	16	0.51%
Failed Procedure	13	0.41%
Fontan Fenestration: Closure by device	12	0.38%
Coronary Interventions	2	0.06%
Grand Total	**3165**	**100.00%**

Table 2. *Radiation data by procedure types. Screening time in minutes, DAP in cGy.cm2f Median effective dose in mSv.*

Procedure Types	Median Screening Time	Median DAP	Median effective dose
Diagnostic Procedure	10.00	59.00	0.76
Atrial communication device occlusion	8.00	578.50	1.63
PDA Occlusion	7.00	136.00	1.44
Pulmonary valvuloplasty	11.00	79.50	1.33
Atrial Septostomy	0.00	0.00	0.00
Coarctation Balloon/Stent/Redilatation	13.00	2022.50	5.42
Pulmonary Artery Angioplasty/stent	28.00	832.15	6.52
PDA Balloon/Stent	3.00	29.90	1.07
Aortic Valvuloplasty	18.00	164.80	2.99
Vascular occlusion by coil/device	26.00	1043.00	7.34
Miscellaneous	17.00	180.65	1.70
Pericardiocentesis	0.00	0.00	0.00
Airway Procedure	4.00	17.00	0.26
Pulmonary vein angioplasty - Stent	37.00	459.55	5.84
RVOT dilatation/Stent	29.00	548.00	6.21
Intervention on systemic-pulmonary shunt	22.00	288.50	5.52
Vascular Access	0.00	0.13	0.00
Paravalvar Leak Occlusion	26.50	9465.00	18.96
Pulmonary Valve RF Perforation	25.00	147.00	5.23
VSD Closure	30.50	2851.25	13.31
Interventional Valve Placement	25.00	3825.50	7.22
Atrial Septal Stent	28.50	138.50	3.51
Failed Procedure	35.00	447.00	4.88
Fontan Fenestration: Closure by device	32.00	3600.00	14.59
Coronary Interventions	42.50	1652.00	9.29

Table 3. Radiation exposure by procedure type, stratified by patient age group when ≥50 cases are available.

Procedure Category	Number	Median Screen Time minutes (IQR)	Median Radiation Dose cGy.cm^2 (IQR)	Median Effective Dose mSv (IQR)
Atrial communication occlusion	**536**	**8.00** (5-12)	**633.3** (186.1-1672.2)	**1.8** (1.0-4.0)
Adult (>18y)	364	8 (5-11)	1100.70 (475-2196)	2.0 (1.0-4.0)
Small Child (2.5y–7.5y)	105	9 (5-12)	140 (63-294)	1.0 (1.0-2.0)
PDA Occlusion	**417**	**7** (4-10)	**139.1** (72-300)	**2.0** (1.0-3.0)
Infant/Toddler (6m–2.5y)	216	6 (4-9)	106.7 (59.6-162.7)	1.3 (1.0-2.0)
Small Child (2.5y-7.5y)	114	6 (4-10)	169.1 (76.5-286)	1.3 (1.0-2.0)
Pulmonary valvuloplasty	**247**	**12** (8-18)	**84** (38-190)	**2.0** (1.0-3.0)
Neonate (0-6m)	131	11.5 (7-20)	52.1 (27.3-104.5)	1.0 (1.0-3.0)
Infant/Toddler (6m-2.5y)	63	12.00 (10-16)	113.1 (63-221)	1.0 (1.0-3.0)
Atrial Septostomy	**219**	**0**	**0**	**0**
Neonate (0-6m)	211	0	0	0
Coarctation Balloon/ Stent/Redilatation	**210**	**13** (10-19)	**2068.5** (599-5476.5)	**5.0** (3.0-11.0)
Adult (>18y)	89	15 (11-21)	4274.5 (2612-7632)	8.0 (5.0-15.0)
Teenager (12.5y-18y)	50	15 (10-18)	2867 (1689-5667)	7.0 (4.0-12.0)
Pulmonary Artery Angioplasty/stent	**195**	**28** (20-43)	**832.2** (261-2118)	**7.0** (3.0-14.0)
Small Child (2.5y-7.5y)	59	29 (20-43)	829.5 (429.3-1113.5)	6.0 (4.0-10.0)
Infant/Toddler (6m-2.5y)	53	24 (17-45)	403 (146-961)	5.0 (2.0-14.0)
PDA Balloon/Stent (all neonates)	**121**	**3** (2-6)	**31.4** (11-82)	**1.0** (0.4-3.0)
Aortic Valvuloplasty	**103**	**19** (13-24)	**181** (75-358)	**2.0** (2.0-5.0)
Neonate (0-6m)	63	19 (13-26)	101 (50-211.6)	3.0 (2.0-6.0)
Vascular occlusion by coil/ device	**93**	**27** (16.75-44)	**1264** (332-4897)	**8.0** (4.0-15.0)

O2568 - ANDRASTENT XL IMPLANTATION FOR THE TREATMENT OF COARCTATION OF THE AORTA IMMEDIATE AND MIDTERM RESULTS

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Introduction: AndraStents are cobalt–chromium balloon expandable, hand mounted stents recently developed for large vessels. We will present our experience on AndraStents implantation for coarctation of aorta (COA).

Methods: Between 2012 and 2016, AndraStent XL were used in 34 patients for COA by hand crimping on BIB, Z-Med or Powerflex balloons.

Results: AndraStents XL were implanted in 34 patients, 23 with native COA and 11 with recurrent coarctation (after balloon angioplasty in seven, and after CP stent implantation in four). Middle aortic syndrome (MAS) was present in two. The median age was 11 years (5–30), and median weight was 38 kg (15–75 kg). Two of them weighed less than 20 kg (15 kg and 17 kg). All procedures were successful (no stent migration, aorta dissection or vessel injury). Two separate stents were implanted for MAS in two and implanted to transvers arc in one. The foreshortening percentages were found to be similar to those claimed by the manufacturer on fluoroscopic images. The vessel diameter was increased from a median 5.4 mm to a median of 12.3 mm, the systolic gradient across the aorta decreased from 35 mmHg before the procedure to 1.5 mmHg after the procedure. In a median 15 months follow up re-dilatation of stents was performed 12 months after only in a patient with MAS.

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Conclusion: AndraStents have several advantages over CP stents: Its material and design allow better tissue penetration and better curve, and a lower crimping profile. It can be used one Fr larger than the balloon shaft in lesions straight ways to reach such as CAO, so it can be used in patients less than 20 kg in weight. A relatively low radial force, tendency to slipping of the stent during the advancing in tortuous way and absence of covered form are disadvantages when compared with CP stents.

O2875 - PERIMEMBRANOUS VENTRICULAR SEPTAL DEFECT DEVICE CLOSURE CHOOSING BETWEEN ADOI AND ADOII

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Background: Transcatheter closure of perimembranous ventricular septal defects (pmVSDs) is a well-established procedure. Recently, Amplatzer duct occluders (ADO) I and II have been reported to close large series of pmVSDs successfully. ADOs are economical compared with the standard Amplatzer VSD occluders; a major consideration in developing countries with low budget programs.
Aim of the Work: We report closure of symptomatic, hemodynamically significant pmVSDs using the ADOI and ADOII devices. Although there are no set criteria for choosing between ADOI and ADOII, the former's price tag includes snare and long sheath. Thus, we aim to predetermine device usage based on transthoracic echocardiography (TTE) findings.
Methods: Between March 2013 and November 2014, 30 patients had transcatheter closure of pmVSDs using the ADO devices. The median age was 4 years (range: 1.1-13 years) and median weight was 15 kg (range: 6.5-85 kg). ADOII could not be used in VSDs larger than 6 mm and/or with a large aneurysm. The median VSD size as assessed by echocardiography was 5.5 mm while the mean was 5.5 mm (range: 3-12 mm), while by angiography it was 5 mm & the mean was 4.75 mm (range: 3-9 mm).
Results: The median fluoroscopy time (FT) was 8 minutes (range: 5 -38 min). We inserted ADOI in 13 patients and ADOII in 17 patients (no significant difference between median age and weight in each group). VSD size was significantly larger and FT was longer in ADOI patients; the device type matched what was decided from TTE data in 84% of cases. Follow up ranged from 2-24 months (median12 months). The mean LVEDD z-score of the patients was 1.1 before VSD closure, while it was 0.63, 0.35 and 0.23 at the 1-month, 3-month and last follow up, respectively. Complete closure rates immediately, at 24 hours and

Table 1. Data for patients who underwent VSD closure.

	ADOI n = 13 (43.3%)	ADOII n = 17 56.7%)	Total n = 30
Sex			
Males	8 (61.5%)	8 (47%)	16 (53.3%)
Females	5 (38.5%)	9 (53%)	14 (46.7%)
Age			
Median (range) in years	5 (1.1-13)	4 (2.3-8)	4 (1.1-13)
Weight			
Median (range) in Kg	17 (6.5-85)	14 (10-45)	
VSD size in mm			
By TTE (mean ± SD)	6.3 (±1.9)	5 (±1.4)	5.6 (±1.75)
By Angio (mean ± SD)	5.87 (±1.7)	4.3 (±0.8)	4.9 (±1.45)
Decision for device as decided by TTE images were matching	84.6%	82%	83.3%
Device size			
10 × 8	9 (69.3%)		
8 × 6	3 (23%)		
12 × 10	1 (7.7%)		
6 × 4		10 (59%)	
5 × 4		3 (17.6%)	
6 × 6		3 (17.6%)	
6 × 5		1 (5.8%)	
Method used to cross the VSD			
Retrograde	8 (61.5%)	14 (82.4%)	22 (73.3%)
Antegrade	5 (38.5%)	3 (17.6%)	8 (26.6%)
Fluoroscopy time (minutes)			
Mean (± SD)	28.47 (±5.64)	22.4 (±8)	27.3 (±8.4)
Z score of LV by TTE (mean (range)			
Before	1 (-0.02 – +4.14)	1.2 (0 – +2.34)	
At 1 month	0.66 (-0.15 – +3.57)	0.59 (-0.82 – +1.8)	
At 3 months	0.41 (-0.71 – +3.57)	0.29 (-0.82 – +1.63)	
At last follow up	0.2 (-0.65 – +3.27)	0.24 (-1 – +1.63)	
Residual VSD	1 (7.7%)	1 (6%)	2 (6.67%)

O2897 - SINGLE CENTER EXPERIENCE OF 65 FETAL AORTIC BALLOON VALVULOPLASTY

Joanna Dangel[1], Adam Koleśnik[2], Marzena Dębska[3], Romuald Dębski[3], Agnieszka Grzyb[2], Grażyna Brzezińska-rajszys[2], Monika Kowalczyk[2], Andrzej Kansy[4], Beata Rebizant[3], Bohdan Maruszewski[4]
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Background: Fetal cardiac interventions(FCI) have been established for changing the natural history of evolving HLHS or rescue fetuses. The national program for FCI started in June 2011, 88 interventions were performed since then.
Material and Methods: We performed 65 fetal aortic valvuloplasty (fBAV) in 60 fetuses. After 10 procedures under general anesthesia we switched to deep mothers' sedation and local anesthesia. Fentanyl and pancuronium was given into the umbilical vein, since 2013 atropine was added. There were 54 fetuses with evolving HLHS and 6 with severe heart failure, 5 hydropic.
Results: 62 procedures were technically successful (95%). In one fetus the valve was not crossed - it was aortic atresia with visible leaflets. In 5 fetuses the procedure was repeated after 1 to 3 weeks. There were 6(10%) fetal demise related to the procedure, 3 of them were hydropic. One fetus developed severe aortic insufficiency after the procedure and died 5 days later. 53 fetuses were live born, 2 of them very prematurely, related to the procedure. There were 3 late premature deliveries by planned CC occurred in different center. Two of those children died, one is on single ventricle pathway. Out of 48 fetuses delivered at term 14 had biventricular circulation (28% from live-born) and alive. 2 did not require any treatment during first 4 months of age. 4 had mitral stenosis with pulmonary hypertension, which made further steps very difficult. 21 alive after single ventricle pathway. There are 70% survivors from all live-born fetuses. Fetuses with border-line left ventricles were the most difficult group for treatment. Hybrid pathway with BV conversion was successful in 2, and failed in 4 fetuses.
Conclusion: fBAV is technically successful procedure, however still there are some unexpected complication. The result of fBAV is unpredictable. The treatment options in neonates and children after fBAV are difficult and complicated.

O2983 - FIRST MULTICENTER TRANSATLANTIC EXPERIENCE WITH THE NEW ULTRASEPT DEVICE FOR ATRIAL SEPTAL DEFECT PERCUTANEOUS CLOSURE.

Jose Diogo Martins[1], Juan Pablo Sandoval[2], Yacine Aggoun[3], Moulin Zinsch[4], Matthias Schulze[5], Matteo Ciuffreda[6], Roberto Mijangos Vázquez[7], Carlos Zabal[2], Fátima F Pinto[8]

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Background: The gold standard treatment for ostium secundum atrial septal defects (ASD) is percutaneous closure. Some concerns remain with the most commonly used devices, namely the inability to access trans-septally the left atrium, the risk of cardiac perforation and niquel content. We present the first-in-man multicenter experience with the 5th generation of the Ultrasept Atrial Septal Occluder Device, released in 2016.
Material and Methods: The Ultrasept is a self-centering, fully retrievable double-disk device that consists of a dual circular design nitinol frame covered with polyvinyl alcohol patches and additional central polyester patch. Its most advantageous features include the extremely low profile, the reduced metal mesh, the septal puncture friendly patch and the flexible and adaptable wire frame. We report a series of consecutive patients from six centers, from Europe and South America.
Results: 44 patients were catheterized with a mean age 11.2 ± 12.7years (min 2.6; max 48.6) and a mean weight 32.0 ± 22Kg (min 7.7; max 94.0). Using transesophageal echocardiogram, the maximal ASD diameter was 8.8 ± 3.1mm (min 5.0; max 15.0) and, with balloon-sizing, it measured 11.4 ± 3.5mm (min 6.0; max 19.5). The procedure was successful in all. The immediate residual shunt was trivial in 14/44, mild in 1/44 and none in the remainder 29/44. No patient showed device interference in venous drainage or valve function. In the 1 month follow-up visit, the closure rate was 100%. Patients were maintained with 6 months of antiaggregation therapy. At 6 months follow up, all patients remain asymptomatic, arrhythmia-free and without residual shunt.
Conclusions: Our current study showed that the new Ultrasept device has comparable short-term safety and efficacy to other commonly used devices and present additional favorable characteristics such as a an extremely low profile, atraumatic and soft device that is trans-septal puncture friendly and with low metal content.

NURSING

O1504 - CULTIVATING EXCELLENCE THROUGH NURSE EMPOWERMENT

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Background: Nurse empowerment can improve patient outcomes through increased nurse retention, high levels of work engagement and innovative behaviors. Empowered nurses experience high motivation, less burnout and job strain, increased job satisfaction, more autonomy and greater respect than nurses in traditionally designed organizations [1–4]. In low-middle income countries, educational and cultural barriers preclude nurse empowerment. The International Quality Improvement Collaborative (IQIC) for Congenital Heart Surgery in Developing World Countries was created in 2007 to reduce mortality through: safe peri-operative practice, infection reduction and team-based practice through nurse empowerment [5].
Materials/Methods: There are currently 47 hospitals from 22 different countries enrolled in the IQIC. Sites participated in monthly didactic webinars designed to facilitate participatory learning, provide a collaborative learning experience, and adapted to individual needs of each center. Infection reduction modules introduced evidence-based nursing practice bundles and specific infection reduction components. Team-based practices focused on standardized communication tools and processes to promote nurse empowerment. Physician-nurse teams from each site oversee project management and quality improvement data collection entered into an IQIC database. Reports are provided to each site with their data and comparison aggregate collaborative data for annual benchmarking. Multidisciplinary IQIC teams conducted site visits for hospital tours, data audits, round table discussions and formal presentations to reinforce collaborative goals.
Results: Since 2010, 25 of the 92 webinars presented advanced nursing content at the request of participating sites. Fifteen nurses from various sites delivered presentations across four IQIC-sponsored international learning sessions. Posters at these sessions described seven, nurse based site initiatives. Two sites have effective nurse-lead infection control initiatives. Risk adjusted mortality and infections have significantly improved at participating IQIC sites [5,6].
Conclusions: Nurse empowerment after IQIC enrollment has been demonstrated by development of nursing leadership, autonomy, high levels of work engagement, innovative initiatives, and improved nurse-sensitive patient outcomes at IQIC sites.

O1516 - HEALTH RELATED QUALITY OF LIFE IN POST OPERATIVE CONGENITAL HEART DISEASE PATIENTS SINGLE CENTRE EXPERIENCE FROM PAKISTAN

Laila Akbar Ladak[1], Babar Hasan[2], Janice Gullick[3], Khadija Awais[4], Ahmed Abdullah[4], Robyn Gallagher[1]
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Introduction: Health related quality of life (HRQOL) issues are common in postoperative Congenital Heart Disease (CHD) patients and often need intervention. There is paucity of data regarding HRQOL in CHD patients from low/middle income countries (LMIC). This mixed methods study conducted in Pakistan aims to provide HRQOL quantitative and qualitative data from CHD patients and their families.
Methods: CHD patients ($n = 140$) and their age matched healthy siblings as controls ($n = 140$), completed the PedsQLTM 4.0 Generic Core and PedsQLTM Cognitive Functioning Scale to determine HRQOL. CHD patients ($n = 22$) and their parents ($n = 20$) were interviewed to explore impact of CHD surgery, challenges and needs after surgery. Matched cohort was compared using t test, predictors were identified utilizing generalized linear model and qualitative data was interpreted through content analysis.
Results: Mean age of CHD patients was 9.98 ± 5.56 years, 68% were male, 50% had a university educated parent and 68% had income below LMIC poverty line. Majority ($n = 98$) had their first

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CHD surgery at >1 year age and mean follow up time was 4.12 ± 1.92 years. Compared to controls, CHD patients had statistically and clinically significant lower HRQOL in all domains, with the worst HRQOL reported for the emotional domain (Table 1). HRQOL was higher in patients with better income, educated parents and an extended family, and lower in those who had longer cardiopulmonary bypass time, higher cardiac prognostic severity, higher RACHS score, re-operations and on cardiac medications. Patients and families highlighted "physical limitations" as their greatest concern; however, "emotional concerns" including "not being normal", "future uncertainty", "impaired family life" and "life-long care" were prevalent and complex.
Conclusions: HRQOL issues effect post-operative CHD patients in Pakistan, with the most disadvantaged being most effected. Solutions need to address the emotional impact on patients and lifelong concerns of parents.

Table 1. Differences in HRQOL – CHD versus Siblings

Domains	CHD Mean ± SD	Siblings Mean ± SD
*Physical	83.77 ± 19.11	98.86 ± 3.35
*Emotional	73.68 ± 17.59	92.46 ± 11.21
*Social	95.00 ± 10.96	99.07 ± 4.74
*School	86.47 ± 17.75	98.48 ± 4.54
*Psychosocial	84.92 ± 12.28	96.09 ± 5.87
*Total	84.66 ± 12.72	96.55 ± 5.31
*Cognitive	90.51 ± 18.27	96.9 ± 9.50

*$p < 0.05$

O1758 - EVALUATION OF CICU ADVANCED PRACTICE PROVIDER EDUCATION AND PRACTICE VARIATION

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Background: The education, training, and scope of practice of CICU advanced practice providers (APPs) is highly variable. A survey was administered to determine specific variations that leave gaps in competence.
Methods: A list of pediatric cardiothoracic surgery programs was generated from the STS database (N = 112). 8 institutions do not have APPs in the CICU/PICU, thus N = 104. A self-administered, electronic survey was delivered via email to 157 APPs at those institutions. 83 APPs responded (53% response rate), representing 36 institutions (35% of institutions).
Results: 65% of respondents started as new graduates. 93-100% obtain a history and physical, order/interpret labs, develop management plans, order/titrate medications and respiratory support. Ability to perform invasive procedures was highly variable but more likely for those in a dedicated CICU. 77% were oriented by another NP, with a duration of orientation <4 months (66%). 83% of APPs with <1 year of experience were oriented by another NP, but 50% of APPs with 7 or more years of experience were oriented by an attending physician. 50% of APPs had no guidelines in place to guide learning/competency during orientation. 67% were not evaluated in any way on their knowledge or skills during or at the end of orientation. Orientation was rated as poor/fair by the majority of respondents for electrophysiology (58%) and echocardiography (69%). 71% rated orientation as moderately effective or less. Respondents stated they would benefit from more structured didactic education with clear objectives, standardized management guidelines, and more simulation/procedural practice. 85% were very/extremely supportive of a standardized CICU APP curriculum.
Conclusions: Orientation for CICU APPs is highly variable, content depends on the institution/preceptor, and competency is not objectively defined or measured. A standardized curriculum is needed and is currently being developed.

O2100 - USE OF HIGH RISK PRECAUTION PROTOCOL TO REDUCE CARDIAC ARRESTS IN THE CARDIAC INTENSIVE CARE UNIT

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Background: Cardiac arrests in pediatric cardiac intensive care units (CICU) are associated with a significant increase in morbidity and mortality. A large multi-institutional cohort estimated overall post-operative arrest rate of 2.6%, arrest rates increased with procedural complexity to as high as 12.7% independent of center volume. Extracorporeal membrane oxygenation (ECMO) is often a rescue intervention during cardiac arrest (eCPR). Survival rate after eCPR is reported just over 50%, with only half of survivors having normal neurological status. Given the vulnerability of this patient population and the negative impact cardiac arrest has on morbidity and mortality rates; our primary aim was to assess the use of a bedside tool highlighting particularly vulnerable patients on arrest rates in the CICU.
Materials and methods: High Risk Precautions (HRP), an inter-professional bedside tool for CICU patients at high risk of cardiac arrest, was created to emphasize high patient acuity, create a framework for improved inter-professional communication, and formally limit nonessential cares and noxious stimuli during this vulnerable period. The tool was generated with medical and nursing team input, trialed, and rolled out to all staff. Patients meeting high risk criteria were placed on HRP.
Results: Forty one tools were collected and analyzed. In the 5 months following implementation of the HRP, we experienced a 38.95% reduction in average monthly rate of cardiac arrests in the CICU, and a 9.39% decrease in the average monthly rate of ECMO deployments. No patients arrested while on high risk precautions. These changes were made despite a high level of new nurse onboarding.
Conclusions: Early data suggest our methods were successful in deceasing arrest rates in the CICU by highlighting this subset of high risk patients, optimizing inter-professional communication regarding clinical goals, and formally limiting nonessential cares and noxious stimuli.

RHEUMATIC

O1244 - ECONOMIC LOAD OF PENICILLIN PROPHYLAXIS ON FAMILIES OF RHEUMATIC HEART DISEASE PATIENTS RESULTS FROM A PEDIATRIC RHD REGISTRY IN INDIA

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Introduction: Rheumatic heart disease (RHD) continues to be a public health problem in developing countries and cost is a major barrier to medication adherence in these regions. The economic impact of penicillin prophylaxis on families of RHD patients is not known.
Methods: We prospectively collected self reported data from the parents of RHD children recruited in the hospital based AIIMS Pediatric RHD Registry. The data included out of pocket expenses on, a) the drug penicillin, b) transportation to the health facility for injection, and c) the provider, for administering injection. We derived monthly estimates of all three categories of costs by adjusting visit costs for frequency of administration. All costs were reported in Indian Rupee (INR), 100 INR are equal to 1.5 US Dollar.
Results: The cost data was provided by 394 patients. The age of patients ranged from 5-18 years (11.5 ± 2.8), 256 (65%) were boys. Most patients were from rural areas (n = 346, 87.8%) and from lower and lower/upper lower socio-economic strata on the modified Kuppuswami socio-economic scale (n = 287, 72.8%). The total monthly out of pocket cost of penicillin prophylaxis was INR 74.7 ± 105.8. The drug cost represented only 33.2% of the total out of pocket costs (INR 24.8 ± 13.7). The cost of transportation to receive the injection (44.6%, INR 33.3 ± 104.9), and the cost of administration (22.2%, INR 16.6 ± 24.2) comprising the remainder of the total cost. The cost of injection provider and the cost of transportation to the health facility exceeded cost of the drug for 37.6% of the patients.
Conclusions: Two third of the total economic load of penicillin prophylaxis on families of RHD patients is constituted by transportation and provider costs. This can be a major barrier to long-term adherence to secondary prophylaxis. Future efforts must be directed at improving access and lowering total cost of secondary prophylaxis.

O1245 - CHANGING THE RHEUMATIC HEART DISEASE EXPERIENCE FOR CHILDREN IN UGANDA THE IMPACT OF A PEDIATRIC SUPPORT GROUP

Amy Scheel[1], *Andrea Beaton*[1], *Emmy Okello*[2], *Chris Longenecker*[3], *Isaac Otim*[4], *Peter Lwabi*[5], *Craig Sable*[1], *Allison Webel*[6], *Twalib Aliku*[7]
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Objective: To assess the impact of a peer-support group to improve knowledge, quality of life, and social support for children diagnosed with rheumatic heart disease (RHD).
Methods: Children from Gulu Municipality diagnosed with RHD and receiving monthly injectable penicillin were invited to participate in a monthly support group for 6 months. Pre- and post-intervention assessments included a baseline RHD knowledge assessment, a measure of pediatric health related quality of life (HRQOL) (PedsQLTM4.0), and a measure of pediatric social support (Hawthorne Friendship Scale). Each session incorporated elements of cooperative play and team building, RHD education, emotional support, and a snack. A short evaluation was used at each session to capture participant experiences.
Results: 42 participants returned parental consent, attended >3 support group sessions, and were included in the data analyses. The majority of participants had latent RHD (83%), identified through previous screenings. Attending support group meetings resulted in increased total pediatric HRQOL scores (60.3 v 70.2, $p < 0.001$), as well as the following pediatric HRQOL sub-scores (physical functioning 55.3 v 68.6 ($p < 0.001$), social functioning 64.2 v 75.8 ($p < 0.001$) and school functioning 59.2 v 69.1 ($p = 0.001$)). Additionally, significant increases in Friendship Scale scores (15.4 v 19.7, $p < 0.001$) and RHD Knowledge scores were observed (3.6 v 6.4, $p < 0.001$).
Conclusions: Peer support groups are effective at normalizing decreased HRQOL scores and increasing RHD disease knowledge and social support. Our findings suggest that peer support groups implemented in conjunction with RHD screening and clinical programs can minimize the negative psychosocial effects associated with early RHD detection.

Table.

	RHD Knowledge		Support Component	
Session	***Topics***	***Supporting material / Activity***	***Activity***	***Discussion starter***
1	N/A	Baseline RHD Knowledge Quiz	QOL and Friendship Scale assessment	N/A
2	Bacteria and strep throat: How one gets RHD and how it can be prevented	Group discussion on myths and fears surrounding RHD	Obstacle course in pairs (one partner is blindfolded led by verbal cues from the other)	Trust: How did it feel to trust someone you just met? Does this relate to your diagnosis with RHD?
3	The heart: anatomy, function and importance	Coloring activity: filling in a black and white image of the heart with red and blue crayons to represent blood flow through the heart	Hula Hoop pass: hold hands and then pass a hula hoop around the circle without unclasping partners hand	Challenges: what obstacles or "hoops" have you had to jump through because of your diagnosis with RHD?
4	The heart: How RHD affects anatomy and function. How RHD is diagnosed	1. Children listened to their own heart with stethoscope 2. Viewing of Echo images of affected Mitral and Aortic valves	Paper Plate Face Mask: create a mask that represents the strongest version of yourself	Feelings: How do you feel when others find out you have RHD? Do they treat you differently?
5	Treatment: the importance of monthly penicillin injections and routine follow ups	Group discussion on myths and fears surrounding penicillin injections	Penicillin Adherence key chains: children spelled out name in beads and then received a heart bead for being in the support group. At each penicillin clinic the children get a new bead to add to the chain	Monthly injections: How do you feel when you have to miss school for injections? Do you tell people where you are going?
6	N/A	Post-intervention RHD knowledge quiz	QOL and Friendship Scale assessment	N/A

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O1889 - EPIDEMIOLOGY AND RISK FACTORS FOR CORONARY ARTERY LESIONS IN CHILDREN WITH INCOMPLETE KAWASAKI DISEASE IN SHANGHAI FROM 2008 THROUGH 2012

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Objective: To analyses the epidemiological features of incomplete Kawasaki disease (IKD) in Shanghai from 2008 through 2012, and investigate the possible risk factors of coronary artery lesions (CAL) in IKD.
Subjects and Methods: This study used the epidemiological survey database of Kawasaki disease in Shanghai from January 2008 through December 2012. The diagnosis of IKD used the AHA diagnostic criteria for KD issued in 2004.
Results: A total of 2304 KD cases were recorded, of which 816 cases were classified as IKD (35.42%). The ratio between male and female was 1.51:1. IKD patients under 1 year of age accounted for the highest proportion (36.40%), and IKD was seen in a highest percentage of patients under 6 months of age (52.69%).There was no statistical difference between CKD and IKD patients in terms of the number of patients receiving IVIG, timing of initial IVIG, the dosage and administration of IVIG. the incidence of IVIG resistance in IKD patients (2.70%) was lower than that in CKD patients (5.71%) (P = 0.001). CAL were found in 90 IKD cases (13.60%) and 201 CKD cases (14.58%) (P = 0.135). The incidence of CAL in IKD infants under 6 months of age (18.36%) was significantly higher than that in other age group (9.01%, P = 0.002), and in these cases, the incidence of CAL was higher than that in CKD patients under 6 m (11.26%, P = 0.026). Risk factors for CAL in IKD patients include age, No. of principle clinical signs, ESR, ALB.
Conclusions: IKD accounts for as high as 35.42% in Shanghai from 2008 through 2012, especially in infants under 6 months, more than half of KD patients presents as incomplete KD. IKD babies less than 6 month have a significantly higher incidence of CAL either than IKD patients in other age groups or than CKD patients in the same age group.

O2036 - THE DIAGNOSIS OF MILD RHEUMATIC HEART DISEASE THREE REVEIWERS ARE BETTER THAN ONE

Elizabeth Tilton[1], Nicola Culliford-Semmens[1], Ross Nicholson[2], John Stirling[1], Rachel Webb[3], Nigel Wilson[1]
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Background: The echocardiographic (echo) features of advanced rheumatic heart disease (RHD) are mostly unequivocally diagnostic of RHD. In contrast, echo screening for RHD predominantly detects milder forms of RHD. For logistical reasons, most screening studies have utilised a single echo reviewer to report echocardiograms. In New Zealand, we have used a 3 reviewer reporting system when practical. The aim of this study was to formally compare the rate of reporting agreement between 2 and 3 reviewers.
Methods: 144 de-identified echocardiograms of children from high prevalence RHD settings were classified by WHF 2012 guideline criteria. Cases included: mild RHD, those with mitral or aortic regurgitation at the upper limit of the physiological range, and congenital valvar abnormalities. Each echocardiogram was independently reported by two reviewers. If there was discordance of the diagnostic category between the two reviewers, a third reviewer reported the echocardiogram. If a 2/3 majority was not reached, an open review of the echocardiogram was undertaken by the three reviewers to reach a consensus (Figure).
Results: The final consensus classifications were Definite RHD (n = 18), borderline RHD (50), upper limit physiological regurgitation/normal (61) and congenital mitral or aortic valve anomaly (15). There was a 60% consensus agreement between the 2 reviewers. There was no significant differences of agreement for the different diagnostic categories, Chi-Squared p = 0.8104. Using 3 reviewers, majority agreement increased to 89%. 11% of cases required an open discussion. (Figure)
Conclusions: The potential variance of one or even two reviewers reporting abnormal echocardiograms of mild RHD is revealed by this study. A three reviewer reporting system is preferable if resources allow.

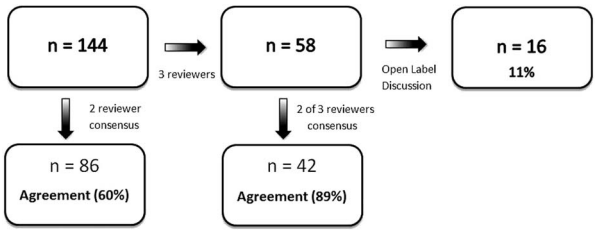

Figure.

O2263 - THE UTILITY OF HIGHLY ABBREVIATED ECHOCARDIOGRAPHIC PROTOCOL FOR DETECTION OF RHEUMATIC HEART DISEASE A PROSPECTIVE POPULATION BASED STUDY IN TIMOR LESTE

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Introduction: Anecdotal evidence suggests a significant burden of Rheumatic Heart Disease (RHD) in Timor-Leste. We conducted the first echocardiography-based screening study to determine the prevalence of RHD in school-aged Timorese children. Additionally, we prospectively tested the utility of an abbreviated echocardiographic screening protocol limited to the parasternal-long-axis-view.
Methods: Students were enrolled from schools in urban and rural districts in Timor-Leste, using opt-out consent that was approved by ethics committees. All students underwent a full screening echocardiogram performed on a Vivid I or Q machine by cardiologists. Following the first parasternal-long-axis-view with 2-D and colour-Doppler, cardiologists were required to log any abnormalities detected prior to proceeding to a full screening study. Those with abnormal screening studies immediately underwent a full congenital echocardiogram. RHD was classified as borderline or definite as per World Heart Federation criteria.

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The prevalence of RHD and the sensitivity and specificity of the abbreviated protocol was calculated.
Results: Total of 1413 children were screened; 739 (52%) female and the median age was 12 years (range 4-24). The point prevalence of definite RHD was 1.8% and borderline 1.6% (total 3.4%). An abnormal parasternal-long-axis-view was documented in 113/1413 (8.0%). No cases of RHD were missed. The sensitivity and the specificity of the abbreviated protocol in detecting RHD was 1.0 and 0.95 (CI 95% 0.94-0.96) respectively. Congenital heart disease was identified in 20 children (1.4%) with 2 needing cardiac surgery. Of these 14 had abnormalities on parasternal-long-axis-view. Only insignificant congenital cardiac defects were missed. All children were linked to care.
Conclusion: This prospective study showed that an abbreviated screening protocol, in school-aged children, limited to a single parasternal-long-axis-view had very high sensitivity and specificity to detect RHD. Only minor congenital defects were missed. Adoption of such screening protocol could make screening more efficacious and practical in regions where RHD remains endemic.

O2425 - ECHOCARIOGRAPHIC SCREENING FOR RHEUMATIC HEART DISEASE IN 4515 SUDANESE SCHOLARS EVIDENCE OF HEALTH INEQUITY.

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Introduction: Rheumatic heart disease is the commonest cause of acquired heart disease in Sudan with a clinical prevalence of 11 per 1000 in 1992.Introduction of hand held echocardiography (HHE) enabled the detection of subclinical disease which was found to be several folds higher than clinical prevalence in many countries. We carried this study to measure the echo prevalence and to initiate a prevention program.
Methodology: This is a prospective epidemiological study on scholars 10-15 yrs of age.phase 1 was conducted in khartoum (september 2015 - Februray 2016). Phase 2: Conducted in south Darfur, Niyala camps for Displaced people (July- september 2016). Echocardiographic (echo) screening using HHE (V scan-GE) was conducted in khartoum by pediatric cardiology fellows using the World Heart Federation criteria. While in Niyala, a simplified protocol using one echo view performed by trained medical officers was utilized.Suspected cases underwent detailed echo examination using standard echo (SE) machine. Training of health personnel was conducted through lectures & workshops.
Results: In Kartoum a total of 3000 scholars were screened,8 cases were found to be positive for RHD by HHE. Out of them,only 1 case was confirmed by SE. The echo prevalance of RHD was 0.3 per 1000. In Darfur, a total of 1515 cases were screened. By HHE, 59 cases were positove for RHD. Outof these 34 underwent SE; 29 of whom were found to have RHD; 22 had definite and 7 had borderline disease. HHE sensitivity was was 85.2%. The estimated prevalence was 33.3 per 1000. A total of 779 health workers were trained and 50 000 posters and pamphlets were distributed. The striking difference of RHD prevalance in 2 areas is an indication of health sevice inequity and the need to mobilize resources to primary health care in remote areas.

SURGERY

O1078 - COMPARISON OF LEFT VENTRICULAR RE TRAINING IN DEXTRO AND LEVO TRANSPOSITION OF THE GREAT ARTERIES

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Background: Patients with both Dextro- and Levo-transposition of the great arteries (D-TGA and L-TGA) are born with the right ventricle (RV) serving as the systemic pump. In both circumstances, the RV may fail as the result of myocardial dysfunction and/or tricuspid regurgitation. Once the RV begins to fail, there are limited medical or surgical options. However, one alternative possibility is to re-train the left ventricle (LV) in preparation for a late arterial switch. The purpose of this study was to evaluate the results of LV re-training in D- and L-TGA.
Methods: This was a retrospective review of 50 patients with D- and L-TGA who were enrolled in an LV re-training program. This included 25 patients with D-TGA, the majority of whom had previously undergone an atrial switch. There were also 25 patients with L-TGA (ie corrected TGA) who did not have a prepared LV due to the absence of a ventricular septal defect or sub-pulmonic stenosis.
Results: The average number of pulmonary artery bands was 1.9 ± 0.4 for D-TGA and 1.5 ± 0.3 for L-TGA ($p < 0.05$). Thirteen of 25 (52%) patients with D-TGA underwent successful LV re-training compared with 24 of 25 (96%) in L-TGA ($p < 0.001$). Ten of 13 (77%) patients with D-TGA underwent successful late arterial switch compared with 18 of 18 (100%) in L-TGA who underwent a double switch procedure ($p < 0.05$).
Conclusions: USIONS: The data demonstrate significant differences in the success of LV re-training and late arterial switch for D- and L-TGA. It is our inference that these differences are related to the condition of the patients at the time of enrollment for LV re-training rather than an inherent difference in LV myocardium.

Table 1. Characteristics of patients enrolled in LV re-training

	D-TGA	L-TGA	p value
Median age (range)	16 years (8 m to 43 y)	11 months (1 m to 24 y)	$p < 0.001$
NYHA class III or IV	23	3	$p < 0.001$
Moderate TR	12	14	p = NS

O1153 - SURGICAL TECHNIQUE FOR ADULT COARTATION REPAIR TWENTY YEARS SINGLE INSTITUTION EXPERIENCE

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Paraplegia following coartaction repair in infants and neonates is a devastating complication.The risk is significantly higher in older children, adults and for recurrent coartaction, with an incidence of up to 5%.In our institution in the last 20 years,99 adult congenital

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patients(>16 years old) were diagnosed with coartation of aorta. All patients were initially considered for an interventional approach and discussed in a multidisciplinary meeting. Sixty-eight adults were treated with either ballooning and/or stenting in the same time period. Thirty-three of them have been treated surgically via median sternotomy or left thoracotomy. Through the surgical approach of left thoracotomy we implemented the use of left heart bypass (LHB) to maintain spinal cord perfusion during repair to provide additional protection and extend the margin of safety and time to execute an accurate repair in more than half cohort. LHB was instituted after full heparinization using centrifugal pump without a reservoir. Blood was returned to the descending aorta distal to the coarctation. The circuit was filled retrogradely with blood from the patient. Femoral blood and left arterial pressure were maintained around a mean of 50 mmHg and 6-9 mmHg. In all our population treated we had only one early mortality, no late mortality and no major neurological/renal complications. There were no abdominal complications. In a mean follow up time of 6.45 years we observed 64% of patients with an improvement of their hypertensive status. In the same follow up time 90% of our patients had at least one MRI scan; only 6% of them showed a mild residual stenosis. In conclusion surgical repair of aortic coartation in adults can be performed safely with excellent early and late results. In addition the use of left heart bypass is an effective technique to augment spinal cord perfusion, and allow careful reconstruction (NB: video would support this presentation).

O1172 - DIAMOND SHAPE ANTERIOR LEAFLET RECONSTRUCTION FOR SEVERE MITRAL REGURGITATION

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Cedimat, Pediatric Cardiac Surgery, Santo Domingo-Dominican Republic[1]; Azienda Ospedaliera Di Padova, Pediatric Cardiac Surgery, Padova-Italy[2]; Children's Hospital of Michigan, Pediatric Cardiac Surgery, Detroit-United States[3]; Cedimat, Pediatric Cariology, Santo Domingo-Dominican Republic[4]; Cedimat, Pediatric Perfusion, Santo Domingo-Dominican Republic[5]; Cedimat, Pediatric Cardiology, Santo Domingo-Dominican Republic[6]; Cedimat, Pediatric Cardiiology, Santo Domingo-Dominican Republic[7]

Background: Repair of severe mitral valve regurgitation due to an anterior leaflet cleft, isolated or in combination with atrioventricular cannal defects, present a challenge and the degree of postoperative regurgitation is the principal risk factor for reoperation and long term survival. We present a technique cleft closure that utilizes a diamond shape patch that restores the normal saddle like anatomy and incorporates native subvalvuvar structures.
Material and Methods: Review of all patients with severe preoperative mitral regurgitation secondary to a mitral cleft that underwent mitral valve reconstruction with diamond shape patch for anterior leaflet cleft reconstruction. Repair was performed with CorMatrix or autologous pericardium. Echocardiographic evaluation of mitral regurgitation was quantified through measurements of indexed vena contracta, related jet and proximal isovelocity surface area (PISA).
Results: From March 2014 to November 2016, 30 patients (mean age 11.6 months , range 4 months to 5 years) with severe mitral regurgitation underwent mitral cleft reconstruction with diamond shape patch using either CorMatrix in 23 (76%) or autologous pericardium in 7 (23%). Included were 16 complete, 8 transitional, 5 partial atrioventricular septal defects, 2 patients with tetralogy of Fallot/atrioventricular septal defect and 2 patients with isolated mitral cleft. There were no early or late deaths. Mean follow up was 9.5 months (range 1 to 24 months). One patient (3.3%) with CorMatrix developed severe mitral regurgitation 6 months after surgery and two (6.6%) with autologous pericardium developed moderate mitral regurgitation 1 and 12 months after surgery. The remaining 27 patients (90%) remain with no or mild mitral regurgitation.
Conclusions: Diamond shape patch cleft reconstruction for severe mitral regurgitation is a simple technique to obtain an anatomical competent valve and avoids artificial chordae implantation in children. Initial results are very encouraging and longer follow up controls.

O1247 - MINIMALLY INVASIVE OFF PUMP TRANSTHORACIC DEVICE CLOSURE OF ISOLATED VENTRICULAR SEPTAL DEFECT 10 YEAR EXPERIENCE

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Objective: Minimally invasive off-pump transthoracic device closure of VSDs under transesophageal echocardiography (TEE) guidance was initialy performed in China in 2007 and has increasingly been performed in China and out of China. The aim of the study was to analyze the safety, efficacy, and long-term follow-up data associated with this technique.
Methods: From March 2007 to November 2017, 632 cases with VSD size from 3 to 12 cm (5.23 ± 4.51 cm) underwent this procedure in our heart center. All the procedures were performed in the operating room. A 2-4 cm inferior sternotomy or the left third intercostals incision was made. Under TEE guidance, through the chosen right ventricle (RV) free wall puncturing point, the delivery system with occlusion device of appropriate size was introduced into RV cavity, defect and then left ventricular (LV) cavity. Then, the LV disc, waist and the RV disc were deployed in LV, septum and RV respectively. After complete TEE assessing for absence of complications, the device was released. Patients were followed up with a standard protocol.
Results: The procedure was successful in 596 cases (94.3%), and was converted to conventional surgical repair in 36 patients (5.7%). The mean device implanting time (puncture to release) was 19.3 ± 12.9 min (3 to 48 min). The mean device size implanted was 6.32 ± 4.25 mm (4 to 14 mm). 565 patients (94.7%) were followed up. During a mean 48.9 ± 26.6 months (1 to 116 months) follow-up period, no mortality occurred. There was no major complication and 54 minor adverse events being detected during the entire follow-up period.
Conclusions: Minimally invasive off-pump transthoracic device closure of VSDs appeared to be excellent with encouraging long-term outcomes. This technique not only provides a third alternative to open heart surgery and transcatheter closure but also shows more advantages.

O1724 - THE ROSS OPERATION IN CHILDREN A TWENTY YEAR SINGLE INSTITUTION EXPERIENCE IN ARGENTINA

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Objective: To analyze the medium and long-term results of the Ross surgery in a young population.
Materials and methods: Between 1995 and 2014, 110 Ross procedures were performed at a median age of 13 years (0.6-19). The aortic valve was incompetent 29.8%, stenotic 10.8% and both lesions were present in 59.4%. A bicuspid aortic valve was documented in 71% of patients (pts). A balloon aortic dilatation was undertaken prior to surgery in 58 pts (64%). The aortic root replacement was the most frequently used surgical technique.
Results: Survival at 5 and 10 years was 94% and 87.5%, respectively. Sixteen reoperations (15%) were needed: autograft replacement 5.4%, RV-PA conduit replacement 5.4% and both 1.3%. The median time between the Ross surgery and the redo was 33.9 months. Free of reoperation at 2 and 5 years was 90% and 78%. The overall mortality rate was 6.3% (7/110). There was a significant decrease in mortality to less than 3% since 2002 and no mortality was registered in the last decade. Incremental risk factors of mortality were: an aorto-pulmonary mismatch >3 mm (n = 4), CPB time >240 minutes (n = 6), ACC time >180 min, age <8 years, weight <20 kg and postoperative arrhythmias (n = 2). From the multivariate analysis, the presence of ventricular dysfunction (n = 6) (OR 7.7 CI95% 0.54-108), postoperative arrhythmias (OR 14.4 CI95% 0.86- 246) were independent predictors of mortality.
Conclusion: The Ross surgery is associated with a good quality of life in the long-term follow-up. A significant aorto-pulmonary mismatch, the presence of postoperative arrhythmias and ventricular dysfunction, as well as a CPB time >240 minutes and ACC time >120 min were associated to higher early mortality. This last one shows a significant decline from 2002 probably associated with the learning curve and the development of ventricular assist devices since 2006.

O1734 - TIMING OF ELECTIVE BIDIRECTIONAL GLENN SHUNT IS EARLY BETTER

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Background: The ideal timing to perform the bidirectional Glenn (BDG) procedure remains unclear. The purpose of this study was to compare the early and late outcomes of patients undergoing a BDG before and after 6 months of age.
Materials and Methods: 550 patients undergoing an elective BDG procedure (1995–2016) were divided into 2 groups: <6 months (n = 200) and >6 months (n = 350). Primary outcomes included operative mortality, survival to Fontan, and survival after Fontan. Data were analyzed using Kaplan-Meier method and log-rank tests.
Results: There was an equal distribution of patients with right and left-sided single-ventricle anatomy between groups. There was no statistical difference in operative mortality [4/200 (2%) vs 2/350 (0.3%), p = 0.20]. Younger patients had a longer hospital length of stay [median 8 days vs 6 days, p < 0.001] and reintervention rate [37% vs 27%, p = 0.02]. Five years following BDG, 338 (61%) patients have undergone Fontan (60%) or BV repair (1%), 37 (7%) have required takedown, transplant or have died, and 175 (32%) were alive waiting (31%) or were not candidates for Fontan (1%). The groups had equivalent interstage attrition [14/200 (7%) vs 22/350 (6%)] (Figure) and hospital survival after Fontan [129/130 (99%) vs 245/253 (98%), p = 0.14].
Conclusions: The BDG procedure is feasible and safe in patients as young as two to three months of age. Despite having longer hospitalization and reinterventions in the interstage period, younger patients demonstrate early and late survival comparable to that seen in older patients.

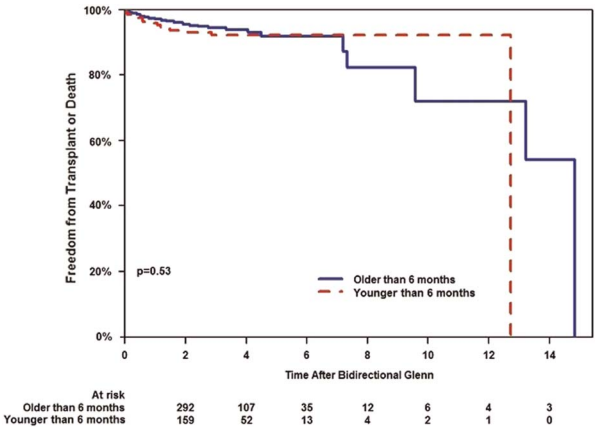

Figure.
Freedom from transplant or death prior to Fontan in patients older and younger than 6 months of age.

O1866 - ISOLATED VS COMPLEX TRANSPOSITION OF THE GREAT ARTERIES – IS THERE STILL A DIFFERENCE IN POSTOPERATIVE PROGNOSIS

Krzysztof Michalak[1], Maciej Moll[2], Katarzyna Sobczak-Budlewska[1], Jadwiga Moll[1], Marek Kopala[2], Tomasz Moszura[1], Paweł Dryżek[1], Jacek Moll[2]
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Background: Arterial switch operation (ASO) is currently the method of choice for transposition of the great arteries (TGA). The additional defects associated with TGA increasing the complexity of ASOs and may have an impact on postoperative outcomes. The aim of this study was to compare the frequency of the most common and severe postoperative adverse events in patients with isolated TGA in reference to the complex cases.
Methods: In this retrospective review, we consecutively enrolled all patients who underwent an ASO for TGA between the years 1991 and 2015. Patients were divided into two group – first one included only those with isolated TGA, without any additional cardiovascular defects, coronary anomalies, with one stage operation performed between 3rd and 14th day of life; the second group included rest of the patients with complex cases of TGA after ASO. We analyzed the difference between those groups in regards to the early and late postoperative mortality, reinterventions (surgical and catheter based procedures), coronary events, significant neoaortic insufficiency (NeoAR) and supravalvular pulmonary stenosis (SVPS).
Results: A total of 715 patients with isolated and complex TGA were included in this study. The average follow-up was 10.5 years (SD:7.6; completeness of the follow-up: 97.3%). The overall early and late postoperative mortality rates were 7.4% and 2.1% respectively, with the significant difference between two analyzed groups (p = 0.025; log-rank test). Freedom from cumulative

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reinterventions at 25 years after the ASO was 92.8% for simple and 82% for complex TGA group ($p < 0.001$; log-rank test). Coronary events were more frequent in patients with complex TGA (8.5% vs 4.9%) but this difference didn't reach the statistical significance ($p = 0.054$;log-rank test). The significant SVPS (>25 mmHg) was associated more frequently with complex TGA (16.1% vs 6.3%; $p < 0.001$; chi-square test). Similarly the presence of significant NeoAR (moderate or severe) was also more frequently associated with complex cases (8.3% vs 4.5%; $p = 0.045$; chi-square test).

O1971 - ARTERIAL SWITCH OPERATION FOR TRANSPOSITION OF GREAT ARTERIES WITH INTACT VENTRICULAR SEPTUM RESULTS OF LATE PRIMARY REPAIR

Domenica Paola Basile, Mauro Lo Rito, Angelo Micheletti, Javier Fernandez Sarabia, Antonio Saracino, Giuliano Giusti, Alessandro Giamberti, Giuseppe Pomè, Alessandro Frigiola, Mario Carminati

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Background: Arterial switch operation (ASO) for transposition of great arteries (TGA) with intact ventricular septum has to be performed in the first weeks of life, to achieve good results and avoid left ventricle deconditioning. The aim of our study is to investigate results of ASO performed after 21 days of life compared to "gold standard" repair performed within the first 21 days of life.
Materials and Methods: Retrospective single center study including all infants with TGA and intact ventricular septum who underwent ASO between May 2004 and February 2016. Patients were grouped according to the age at the operation: "early repair" (age < 21 days) and "late repair" (age > 21 days). Differences among groups were addressed in terms of postoperative outcomes: in-hospital mortality, incidence of ECMO and left ventricular dysfunction.
Result: Infants included in the study were 137 (male = 93). 55 cases (40%) were in the "late repair" group. Pre-operative echocardiogram showed "banana shaped" left ventricle in 38 patients; 35 out of 38 belong to the "late repair" group. Yacoub type A (n = 91) and Yacoub type D (n = 31) were the most common coronary patterns detected. Intramural course was found in 10 cases. Overall hospital mortality was 8% (n = 11) with no statistically significant difference between the two groups (late = 5% Vs. early = 10%, $p = 0.364$). ECMO incidence in late group (13%) is almost double compared to early repair group (7.5%) although not statistically significant ($p = 0.295$). 6 patients had post-operative left ventricular dysfunction at discharge with no differences in incidence among groups ($p = 0.536$). Median hospital stay was similar between groups (late = 22 days Vs. early = 20 days, $p = 0.223$).
Conclusions: Primary late ASO has led to similar results compared with early ASO although the need of post-operative ECMO, to support left ventricle recovery, remains higher in this group. Primary ASO can be performed, with low in-hospital mortality, in infants more than 21 days.

Table.

Patient's variables	Total	Early repair (n = 82 pts) N (%)	Late repair (n = 55 pts)	P-value
Sex (male)	93	58 (70%)	35 (68%)	0.383
Coronary pattern				
Type A>	91 (66%)	54 (66%)	37 (67%)	0.634
Type B	4 (2%)	2 (2%)	2 (4%)	
TypeC	7 (5%)	6 (8%)	1 (2%)	
Type D	31 (23%)	18 (22%)	13 (23%)	
TypeE	3 (2%)	2 (2%)	1 (2%)	
Intramural	10 (7%)	8 (10%)	2 (3.6%)	0.177
Banana shape	38 (28%)	3 (4%)	35 (64%)	0.000
Age at surgery - days (median, IQR)	8.5 (7-12)	27 (25-36)	13 (8-27)	0.000
Cardiopulmonary bypass time (min) (median, IQR)	150 (125-175)	145 (122-179)	150 (134-165)	0.7812
Aortic cross clamp time (min) (median, IQR)	90 (75-108)	75 (70-90)	85 (125-175)	0.022
Chest left open (days) (median, IQR)	3 (2-4)	3 (2-3)	3 (3-5.5)	0.0337
ECMO support	13 (10%)	6 (7.5%)	7 (13%)	0.295
Hospital mortality	11 (8%)	8 (10%)	3 (5%)	0.364

O2042 - SUCCESSFUL MANAGEMENT STRATEGIES FOR THE HIGH RISK FONTAN IN THE SETTING OF HYPOPLASTIC LEFT HEART SYNDROME

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Objective: Patients at high risk for Fontan completion include those with poor haemodynamics and unfavourable anatomy. This a particular concern in Hypoplastic Left Heart Syndrome (HLHS) with pulmonary vascular problems or impaired ventricular function. We identified successful management strategies and risk factors for Fontan completion in patients with HLHS in order to define high risk patient groups and examine outcomes in relation to decision making and preoperative preparation.
Methods: All HLHS patients assessed for Fontan completion in a single centre from 2005-2016. High-risk classified as: (1) Poorly developed pulmonary vasculature; (2) High pulmonary Vascular Resistance (PAP > 15 mmHg + /-TPG > 6 mmHg); (3) ≥ moderately impaired ventricular function and end diastolic pressure > 14 mmHg. Outcomes analysed with focus on early Fontan failure (defined as takedown, prolonged ICU stay/chest drainage, or transplantation/death).
Results: Of 225 HLHS patients assessed, 201 underwent Fontan completion. Thirteen(6%) were declined due to poor pulmonary vasculature/lymphangectasia (n = 3) or severely impaired ventricular function (n = 10). The latter were all referred for transplantation and 4 died. No deaths were reported at 30 days post Fontan, and 4 deaths (2%) between 60 days and 6 years(median follow-up of 5.0 years). Three patients required Fontan takedown and two transplantation. Seventeen (8%) had raised PVR pre-operatively and were treated with sildenafil. Fifteen responded and underwent successful Fontan; 5 patients continuing on sildenafil long term. Two patients with impaired function received preop dobutamine infusion before successful Fontan. Patients who failed Fontan had relatively low mean Nakata Index ($195 \pm 31 mm^2/m^2$). Preop PAP and TPG were not associated with Fontan failure. No other risk factors were identified for prolonged pleural drainage although there was a trend for smaller Nakata Index.
Conclusion: Careful patient selection can deliver good Fontan outcomes in the HLHS group. Interim treatment with sildenafil is a useful adjunct in patients with high PVR. Poorly developed pulmonary vasculature is associated with later Fontan failure.

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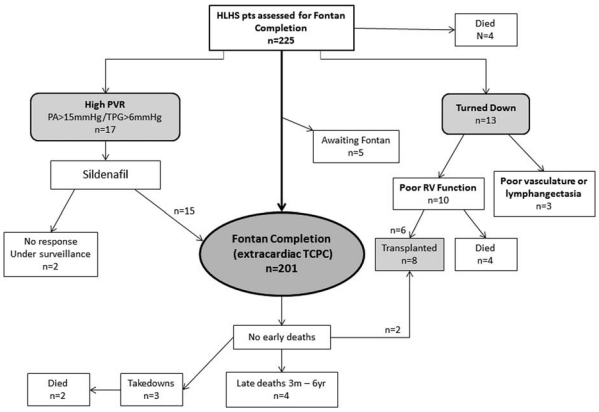

Figure.

O2093 - RISK FOR AND OUTCOMES OF PERI OPERATIVE STROKE ASSOCIATED WITH THE FONTAN PROCEDURE

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University of Alberta, Surgery, Edmonton-Canada[1]; University of Alberta, Pediatrics, Edmonton-Canada[2]; University of Alberta, Radiology, Edmonton-Canada[3]; University of Alberta, Critical Care, Edmonton-Canada[4]

Background: Studies looking at risk of stroke in single ventricle patients have focused on long-term outcomes, with little specific attention paid to the peri-operative period. We aimed to assess predictors of stroke at the time of Fontan operation.
Methods: All children who underwent a Fontan operation between 1996 and 2016 were included. Data including anatomy, surgical technique, and cardiopulmonary bypass details at Fontan were obtained from the surgical database and patient charts. Peri-operative stroke was defined as any clinically evident cerebrovascular event during the Fontan hospitalization. Characteristics were compared between subjects who suffered a stroke and those who did not; the association between stroke and length of stay (LOS) and mortality was also evaluated.
Results: This study included 320 subjects (107 hypoplastic left heart syndrome (HLHS), 135 female), 23 (7.2%) of whom suffered a perioperative stroke. Confirmatory neuroimaging was available for 20 (87%) of the stroke subjects. There were 17 deaths (6 peri-operative) over the course of follow-up; 10-year survival was 95.2%. Median age (3.3 vs 3.3 years) and weight at Fontan (14.2 vs 14.1 kg) did not differ between groups, nor did Fontan type, use of fenestration, or cardiopulmonary bypass times (all $p > 0.05$). However, subjects with HLHS were more likely to suffer a stroke than those without (12.2% vs 4.7%, $p = 0.015$), as were children who had part of their procedure done with fibrillatory arrest (16.7% vs 5.5%, $p = 0.006$); these associations persisted in multivariate analysis. A peri-operative stroke was associated with a longer LOS (median 20 vs 10 days, $p = 0.0001$), but not mortality: one child died post stroke, after having left hospital (HR 1.23, 95% CI 0.29, 5.23).
Conclusions: Peri-operative stroke is not uncommon with the Fontan operation, especially in children with HLHS anatomy. These children do survive but likely face challenges in terms of education and quality of life.

O2109 - AUGMENTATION OF THE PULMONARY ARTERIES IS NOT ASSOCIATED WITH WORSE LONGER TERM OUTCOME A PROPENSITY MATCHED ANALYSIS FROM THE AUSTRALIA AND NEW ZEALAND FONTAN REGISTRY

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Lady Cilento Childrens Hospital, Queensland Paediatric Cardiac Service, Brisbane-Australia[1]; The Childrens Hospital At Westmead, Heart Center For Children, Sydney-Australia[2]; Princess Margaret Hospital For Children, Childrens Cardiac Centre, Perth-Australia[3]; The Womens and Childrens Hospital, Paediatric Cardiology, Adelaide-Australia[4]; Starship Childrens Hospital, Green Lane Paediatric and Congenital Cardiac Service, Auckland-New Zealand[5]; Royal Childrens Hospital, Cardiac Surgery, Melbourne-Australia[6]

Background: Adequacy of the pulmonary arteries is essential for a durable Fontan circulation. Long-term data following pulmonary artery (PA) augmentation in association with the Fontan is lacking.
Methods: Pre-operative, operative and follow-up data for 1437 patients in the Australia and New Zealand Fontan Registry (1975-2015) was analysed. The primary endpoints were death and death or failure (takedown, transplant, PLE or plastic bronchitis). Cox regression with propensity score matching (1:1 nearest neighbour) was used to determine the risk or benefit conferred due to pulmonary artery augmentation.
Results: 48 (3.3%) patients underwent PA augmentation after a cavo-pulmonary shunt but prior to Fontan (surgical n = 14, balloon dilatation n = 16, stent insertion n = 19). 12 of these patients required further intervention at the time of Fontan (surgical = 7, intraoperative stent = 4, combined = 1). A further 105 (7.6%) patients underwent PA augmentation at the time of Fontan (surgical = 104, stent = 1). Median follow-up was 6.4 years with 10 deaths (6.5%) in the PA augmentation group and 10.5 years with 95 deaths (7.4%) in the non-augmentation group. Tests for frailty showed no significant variance in outcome between surgical centres or different decades. Initial unadjusted Kaplan-Meier log-rank and Cox regression demonstrated no significant difference in both endpoints between the 2 groups (death: HR 1.35, 95% CI 0.70–2.60, $p = 0.366$; death or failure: HR 1.39, 95% CI 0.83–2.34, $p = 0.211$). Propensity score matching yielded a total of 262 matched patients (131 in each group) with adequate balance for all covariates (median residual bias = 0.05). Subsequent Cox regression demonstrated no significant difference in the risks of death (HR 1.30, 95% CI 0.49–3.41, $p = 0.596$) and death or failure (HR 0.92, 95% CI 0.46–1.85, $p = 0.817$)
Conclusion: The requirement for PA augmentation prior to or at Fontan does not confer worse longer-term outcomes. An aggressive approach towards PA augmentation for hypoplastic pulmonary arteries is justified in prospective Fontan patients.

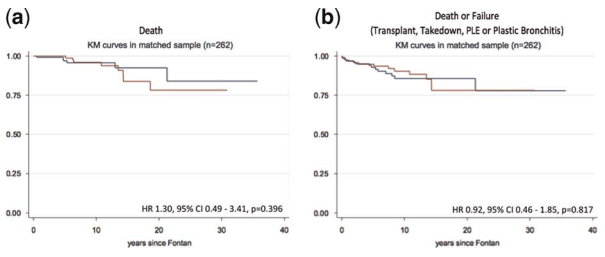

Figure 1.
Kaplan-Meier (KM) Survival Curves after Fontan completion for a) death and b) death or failure (Transplant, Takedown, Protein-Losing Enteropathy (PLE) or Plastic Bronchitis). Red = Pulmonary Artery Rehabilitation, Blue = propensity matched controls.

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O2145 - CLINICAL OUTCOME OF CONTEMPORARY EXTRACARDIAC CONDUIT TOTAL CAVOPULMONARY CONNECTION A 15 YEAR SINGLE CENTER EXPERIENCE

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Objectives: This study aimed to evaluate the clinical outcome of contemporary extracardiac conduit total cavopulmonary connection and identify risk factors affecting early and late outcomes.
Methods: Between January 2000 and December 2014, 453 patients (median age, 6.7 years, range: 2.1 to 36.6 years) with a wide range of underlying diagnoses underwent ECC with a prior (n = 202) or simultaneous (n = 251) bidirectional cavopulmonary shunt. Thirty six patients (7.9%) showed heterotaxy, and fenestration was created in 247 patients (54.5%). The clinical outcome were retrospectively reviewed from the medical records. The median follow up was 4.5 years with a maximum of 15.5 years. Survival was examined with the Kaplan–Meier method, and risk factors were analyzed in the multivariate hazard domain.
Results: There were 21 early and 8 late deaths. The Kaplan–Meier estimated survival rate was 94.4 ± 1.1% at 5 years and 88.7 ± 3.7% at 10 and 15 years. Late-occurring morbidities included tachyarrhythmia in 10, bradyarrhythmia in 4, protein losing enteropathy in 5 and thromboembolism in 7 patients. Freedom from late occurring morbidities at 15 years was 82.6% ± 5.1. Outcomes were equivalent between fenestrated and nonfenestrated patients. Among survivors, 95.4% were classified in New York Heart Association class I or II at the latest follow-up. Multivariable analysis demonstrated heterotaxy, right ventricular morphology and longer cardiopulmonary bypass time to be independent risk factors for early death. Multivariable Cox regression revealed heterotaxy (odds ratio [OR]: 4.504, p = 0.001) and longer cardiopulmonary bypass time (OR: 1.009, P < 0.001) as risk factors for death, and elevated preoperative pulmonary artery pressures (P = 0.013) as risk factor for Fontan complications.
Conclusions: Contemporary extracardiac conduit total cavopulmonary connection can be performed with low mortality and morbidity rates and achieves excellent long-term outcomes. Heterotaxy and longer cardiopulmonary bypass time was associated with death.

O2153 - MID TERM OUTCOMES OF AORTIC ARCH RECONSTRUCTION IN NEWBORNS AND INFANTS A 5 YEAR EXPERIENCE FROM A START UP CONGENITAL HEART SURGERY PROGRAM

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Background: The aim of the study was to present our experience in aortic arch reconstructions in newborns and infants with or without concomitant pathologies.
Materials and Methods: Between October 2011 and December 2016, 96 newborns and infants with hypoplastic aortic arch with or without concomitant pathologies underwent aortic arch reconstruction as primary operation. Patients were retrospectively analyzed using the hospital database. The demographic data, additional procedures and clinical outcomes were reviewed. Risk adjustment analysis was performed using the Aristotle basic scores (ABC), Aristotle comprehensive scores (ACC) and Complexity levels (CL).
Results: Seventy-seven (80%) patients presented with two-ventricle physiology and the others (20%) had single ventricle. All patients (male/female: 58/39) were operated through median sternotomy with cardiopulmonary bypass. Mean age was 72 days (range: 3- 360). Mean weight was 4,1 kg (range: 2,2-9,2). A high correlation was found between the ABC, ACC and CL and mortality ($p < 0.05$), indices of morbidity and technique difficulty (Table1). Fourteen (14.5%) patients needed extra corporeal membrane oxygenation, 4 of them were discharged and followed-up. In hospital mortality developed in 20 cases with significantly higher risk scores and complex additional procedures. No late mortality occurred during mean follow-up time of 477 days (range: 20-1800). During follow-up, 8 (10.5%) of 76 patients need a re-intervention for re-coarctation, which included re-operation (n = 3) and balloon dilatation (n = 5).
Conclusion: In patients who undergo surgery for aortic arch hypoplasia, postoperative morbidity and survival seems to be more dependent on preoperative characteristics and ABC, ACC and CL than the type of surgery.

Table 1. Aristotle risk scores of patients.

	ABC	ACC	CL
Isolated aortic arch surgery[@]	7	7	2
All patients	7.9	10	2.5
Survivors	7.7	9.5	2.4
Non-survivors	8.9	12.2	3.1

Data are presented as the mean values. Note that risk scores of non-survivors are more than survivors ($p < 0.05$).
[@] = expected risk score in isolated arch surgery in Aristotle scoring system. ABOAristotle basic score, ACC = Aristotle comprehensive score, CL = complexity levels.

O2160 - LONG TERM RESULTS AFTER SURGICAL REPAIR OF AVSD OVER A 30 YEARS' PERIOD AT A SINGLE CENTRE INSTITUTION

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Background: Despite improved survival after atrioventricular septal defect (AVSD) repair over the last decades patients are still at risk for reoperation especially addressing left atrioventricular valve. We analyzed our 30 years' experience to define risk factors for mortality and reoperation after AVSD repair.
Methods: Between 1986 and 2016 532 patients received AVSD repair. 2.4% of these patients had unbalanced AVSD, but biventricular repair was feasible. Complete AVSD was presented in 72.0%, intermediate AVSD in 21.7% and partial AVSD in 6.3%. Median age was 6.2 months, 50 patients were younger than 3.5 months. Standard AVSD repair was performed using two patch technique (81.5%) and cleft closure (97.3%). 56 patients received previous surgery (72% pulmonary artery banding).

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Distribution of trisomy 21 was 59.4%. Results were divided into two surgical eras (1986-1999 and 2000-2016).
Results: Surgical era strongly correlated with mortality: In-hospital mortality decreased from 9.9% in pre-millennium era to 2.6% in post-millennium era ($p < 0.001$). Median follow-up time was 14.3 years. Freedom from reoperation was 79.7%. Main indication for reoperation was left ventricular valve regurgitation. 13.5% received mitral valve repair, 6.2% mitral valve replacement. Previous mitral valve repair was associated with mitral valve replacement. 4.2% received reoperation for left ventricular outflow tract obstruction and 4.3% for tricuspid valve regurgitation. Reoperation rate was significantly lower in patients with trisomy 21 ($p < 0.001$). Age lower than 3.5 months correlated with mortality and reoperation, especially for mitral valve repair ($p < 0.001$). Mortality and reoperation rate did not differ between patients with unbalanced AVSD and balanced AVSD.
Conclusion: AVSD repair can be performed with excellent results and good long term survival. Patients with trisomy 21 demonstrate no difference in survival but lower reoperation rates. Left atrioventricular valve regurgitation is main indication for reoperation.

O2259 - EARLY OUTCOME OF FONTAN OPERATION BASED ON AGE EXPERIENCE OF TERTIARY CARDIOVASCULAR CARE IN INDONESIA

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Background: Fontan procedure represents the final stage for the palliation of univentricular heart and early intervention for optimal result is important. In developing countries, patients mostly came late due to many problems and outcomes after Fontan operation is then a matter of concern.
Objective: This study aimed to evaluate the clinical outcome of intra hospital and 1 month after Fontan operation at National Cardiovascular Center Harapan Kita, as the tertiary care in Indonesia.
Method: We reviewed patients underwent Fontan operation between 2006 and 2016. Patients were divided into two groups, group A less than 5 years old ($n = 70$), and group B above 5 years of age ($n = 78$). Pre-operative, operative and post-operative data and 1 month outcomes were assessed.
Results: Total of 148 patients old with median age at operation 6 (3-22) years were included. Hospital mortality was 8 (11.4%) in group A and 7 (9%) in group B ($p = 0.621$). Pleural effusion duration and length of hospital stay were not significantly different between two groups 14.9 ± 2.26 vs $12.36 \pm 10{,}64$ days, $p = 0.39$, and 20.61 ± 13.4 vs. 22.13 ± 17.4 days, $p = 0.657$, respectively). Re-hospitalization within 1 month post-operative course found in 9 (12.9%) patients group A and 15 (19.2%) patients group B ($p = 0.294$). Re-hospitalization which mostly due to recurrent pleural effusion (protein loosing enteropathy) was significantly higher in older group ($p = 0.016$). Re-operation were also not significantly different between both groups (1 (1.4%) vs. 5 (6.4%), $p = 0.125$).
Conclusions: There were no differences in morbidity, mortality, and re-hospitalization between age groups of Fontan operations. Rehospitalization caused by recurrent pleural effusion due to protein loosing enteropathy was more frequent in older group.

O2320 - TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION IMPROVED EARLY OUTCOMES WITH AN ALTERNATE MANAGEMENT STRATEGY

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Background: Surgical repair of TAPVC has been associated with significant mortality especially in developing countries, largely blamed on post-operative pulmonary hypertensive crises. We believe that altered ventricular dynamics post-operatively are more responsible for mortality and have evolved a management strategy accordingly. The results of this strategy are presented.
Patients and Methods: 283 patients (median age -93 days; median weight -4100 gms) with isolated TAPVC underwent repair between Jan 2001 and Dec 2016. 51% had supracardiac, 27% had cardiac, 10% had infracardiac and 12% had mixed connections. 140 (49.5%) had pulmonary venous obstruction at some level. 17% required emergent surgery, 24% were semi-urgent and the rest were done electively on priority. Operative strategies included 1) normothermic bypass with short cross-clamp duration 2) posterior approach for supra and infra cardiac connections 3) routine ligation of the vertical vein. Post-operative strategies included 1) routine left atrial pressure (LAP) monitoring instead of pulmonary artery pressure monitoring to track left ventricular function 2) aggressive afterload reduction 3) frequent echocardiographic assessment of bi-ventricular dynamics and 4) weaning from ventilator support only when LAP was consistently low.
Results: In-hospital mortality was 5/283 (1.76%). Causes of death were severe sepsis -3, bronchomalacia -1, malignant arrhythmia-1. No patient required mechanical support or inhaled nitric oxide. Median ventilator duration and hospital stay was 52 hours and 11.2 days respectively. All survivors were discharged with documented unobstructed pulmonary venous drainage.
Conclusion: Early outcomes following surgery for TAPVC can be improved by a strategy focused on tracking ventricular function rather than pulmonary artery pressure. Nitric oxide and mechanical support are easily avoidable.

O2499 - LEFT VENTRICLE RETRAINNING OF D TRANSPOSITION OF GREAT ARTERIES IN THE THIRD WORLD. THE BUCARAMANGA EXPERIENCE

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Introduction: A left ventricle (LV) dysfunction is a usual natural development frequently found in our country, regarding to a delayed diagnosis of D-Transposition of Great Arteries (DTGA) in infants. In this scenario Pulmonary Arteries (PA) banding was performed to train the LV for anatomical repair.
Methods: LV retraining was performed in 18 patients with DTGA intact ventricular septum (IVS) from 2006 to 2016,

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when a "Banana Shaped" LV and/or LV mass <35 gr/m2 was diagnosticated.

Results: The mean age for retraining was 60 days ranging from 24 days to 4 years. Mean weight 4,3 kg (3.5-11 kg) (table1). Causes of LV dysfunction were late diagnosis, sepsis, neurologic complications and a patient with LV dysfunction since birth. Pre-training Mean LV mass was 28 gr/m^2 with a gain of 2.5 gr/m^2 in the first 24 hours, 14 gr/m^2 (10days), 30 gr/m^2 pre-switch. Pre-switch LV mass was not related with ECMO post-switch (p 0.84). There was no relation between the patients in the 75 percentile of mass gain and age or weight pre-training. Initially 72% had a grade III deviation of ventricular septum, 28% grade II. Pre-switch 46% grade I, 54% grade II. 39% changed from Grade III to II immediately post banding. Two patients died from cardiac failure, another two from non-cardiac causes. Three patients needed ECMO after arterial switch with 2 survivors. There was no mortality during the LV retraining period. Five patients are pending the arterial switch operation.

There was a 78% post- arterial switch survival.

Conclusions: Although controversial, LV mass and septum morphology still are simple and useful tools in LV dysfunction analysis. LV retraining is a viable option for patients with LV dysfunction planned for an anatomical repair of DTGA with IVS, in patients once considered not suitable for a direct switch procedure in our country.

Table.

Age		
Median	75	
RIQ	52-390	
Rank	24-1460	
Gendre (Male)	15	83%
Weight to preparation		
Median	4,4	
RIQ	3.7-5.9	
Rank	3.5-11	
Cause		
Late diagnosis	10	56%
Neurologic dispairement_ Hipoxia	4	22%
Late remission	**1**	6%
Sepsis	1	6%
low weight	1	6%
De-functionalised from birth	1	6%
Time from preparation to switch (days)		
Median	49	
RIQ	23-56	
Rank	7-993	
Age at switch		
Median	153	
RIQ	109-457	
Rank	39-2453	
Ventricular morphology		
Grade II	5	28%
Grade III	13	72%
General survival	14	78%
Cardiac survival	16	89%
Venricular mass before preparation		
Median	28	
RIQ	19-33	
Rank	7-53	
FEVI before pre- preparation {%)		
Median	74	
RIQ	62-80	
Rank	50-90	

O2556 - CONTINOUS METABOLIC MONITORING ALLOWS TAPERING OF NORMOTHERMIC AND HYPOTHERMIC CARDIOPULMONARY BYPASS DURING OPEN HEART REPAIR IN INFANCY

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Backgroound: Traditionally, perfusion parameters for cardiopulmonary bypass (CPB) in infants have been based on body weight, resulting arguably in overestimated nominal flows related with higher morbidity. Aim of the study was to assess safety and efficacy of a CPB strategy that implemented continuous metabolic and hemodynamic monitoring during open-heart repair in infants.

Materials and Methods: 31 patients undergoing surgery using normothermic CPB (mean age: 1.4 ± 1.7 years, mean body weight: 7.8 ± 3.8 kg) and 23 patients using hypothermic (25 °C) CPB were enrolled (mean age: 9.4 ± 24.0 months, mean body weight: 4.8 ± 3.5 kg). The patients were managed according to conventional CPB flows (150 mL/min/kg), except for a 20 minutes test during which CPB was adjusted to the minimum flow to maintain MVO2 >70% and rSO2 >45%. Hemodynamic, metabolic and clinical variables (along with gaseous microemboli (GME) generation in a subgroup of twenty patients) were compared within patient groups. Moreover, in a recent pilot study we evaluated the impact of metabolic guided CPB on renal function assessing splanchnic NIRS and post-operative urine output and creatininemia trend.

Results: The 20 minute test allowed reduction of CPB flows greater than 10% and 20% ($p < 0.05$) respectively in the normothermic and in the hypothermic CPB population, with no impact on pH, blood gas exchange and lactate. There was 1 (1.8%) in-hospital mortality, no neurological morbidity and no need for ECMO support. Moreover, mean GME generation was greatly decreased during the metabolic-guided CPB test, standing at 15% and 45% ($p < 0.05$) of the value recorded during standard CPB, respectively on the arterial and venous line. Preliminary results from the pilot study seem to suggest no major impact on renal function.

Conclusions: The present study shows that a safe individualized strategy of conduction of CPB, which allows significant flow reduction, maintaining normal haemodynamic and metabolic parameters, both in normothermia and moderate hypothermia may be possible.

O2602 - INTRAOPERATIVE REVISION DURING ARTERIAL SWITCH OPERATION IN THE CURRENT ERA

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Background: Arterial switch operation is a relatively safe operation nowadays. We hypothesize that intraoperative revision for coronary malperfusion still plays an important role in the current era.

Methods: From January 2005 to December 2016, 255 patients underwent arterial switch operation. Factors related to the need for intraoperative revision were analyzed. Morbidity was compared in patients with/without intraoperative revision. A subanalysis was performed for 134 patients operated on by the most senior surgeon, and the "standard technique" for a simple anatomy

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and "modified technique" for a more complex anatomy were defined. The factors related to the application of the "modified technique" and the relationship between "modified technique" and intraoperative revision were analyzed.
Results: Seventeen patients (7%) needed intraoperative revision, and factors related to intraoperative revision were body weight at surgery ($p = 0.051$) and eccentric position of the coronary ostium ($p = 0.01$). Discharge mortality was 2% overall: 2/17 with intraoperative revision vs 3/238 without it ($p = 0.04$). Patients who underwent intraoperative revision had longer postoperative hospitalization ($p = 0.003$). Application of the "modified technique" was related to unusual coronary artery origin and branching pattern ($p < 0.001$), major malalignment of the commissures ($p < 0.001$), and eccentric position of the coronary ostium ($p = 0.007$). Application of the "modified technique" did not correlate with intraoperative revision ($p = 0.51$).
Conclusions: The need for intraoperative revision was related to eccentric coronary ostium but not to unusual coronary artery origin and branching pattern. Although the need for intraoperative revision was related to higher mortality and morbidity, it likely also contributed to our excellent results after arterial switch operation.

O2638 - IS BILATERAL GLENN A RISK FACTOR PRIOR TO FONTAN COMPLETION

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Objective: Safe Fontan completion requires normal ventricular function, pulmonary arteries (PA) anatomy and low pulmonary vascular resistances (PVR). Bilateral Glenn (BBDG) is generally associated with PA bifurcation stenosis and presents multiple sources of pulmonary blood flow. We suggest that in BBDG prior to Fontan, PVR are underestimated and ends up with a less optimal outcome.
Methods: From 2008 to 2016, 98 patients with single ventricle anatomy enter a Fontan pathway. 80 had a single superior Vena Cava (SVC): G1 whereas 18 had 2: G2. Both groups were similar in terms of demographic, intra cardiac anatomy, number of previous surgeries and physiologic functional data prior to Fontan procedure. All received extra cardiac Fontan, 53% with fenestration. Uni and multivariate analysis were performed with regard to mortality and failure. A p-value <0.05 was considered as significant.
Results: There were more heterotaxy in G2. BBDG gave better growth of both right and left PA but created a PA bifurcation stenosis. Stormy post operative course with elevated liver enzymes and renal failure, mortality rate (22% vs 3.75%) and Fontan failure rate (21.5% vs 6.5%) were higher in G2. Multivariate analysis showed that heterotaxy ($p = 0.027$) and bigger size of right PA ($p = 0.035$) only were independant risk factors for poor outcome, the latter meaning that pulmonary flow distibution is uneven. Survival and Freedom from Fontan Failure rates were 93.5% and 87% in G1 vs 78% and 72% in G2 (p = ns) at 5 years post operatively.
Conclusions: This study demonstrates that BBDG present multiple sources of pulmonary blood flow with a stenosis in between meaning separate right and left lungs blood flows. Calculation of PVR prior to Fontan, should be performed in accordance with this anatomo physiologic consideration.It also suggests that surgical technique should address this anatomy either early in life or at time of Stage II.

O2762 - AORTIC VALVE RECONSTRUCTION WITH THE OZAKI PROCEDURE IN PEDIATRIC PATIENTS EARLY OUTCOMES IN 21 PATIENTS

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Introduction: The Ozaki procedure with complete cusp replacement has shown excellent results in the adult population. Early results with pediatric patients with congenital aortic valve disease are reported here.
Methods: A retrospective analysis of aortic valve reconstruction using the Ozaki procedure was performed on 21 consecutive patients (15 male) with congenital aortic valve disease at a single institution from March 2015 to September 2016. Outcome measures included mortality, surgical or catheter-based re-interventions and echocardiographic measurements.
Results: Eleven patients had AR and 10 patients AS/AR. Seven, 11 and 3 patients had tricuspid, bicuspid and unicusp valves, respectively. Eleven patients had a prior aortic valve repair and one patient had a prior bioprosthetic AVR. Median age at surgery was 10.5 years (IQR 5-14) and weight was 27.4 kg (IQR 18-60). There were 2 infants (1-2 years), 13 children (3-12 years), 4 adolescents (13-17 years) and 2 adults (ages 23 and 24). Pre-operative echocardiography revealed the average annular diameter was 2.07 ± 0.44 mm and peak gradient for AS/AR patients was 57.18 ± 21.14 mmHg. Three leaflets replaced in 15 patients, two in one patient, and one in five patients. Autologous and photofix bovine pericardium were used in 11 and 10 patients. There were no mortalities or conversions to valve replacement. Median hospital and ICU LOS was 5.25 and 1.87 days. Median follow-up was 4 months (range, 1-13). At discharge, no patients had more than mild regurgitation and mean peak gradient was 14.15 ± 8.35 mmHg. At late follow-up (5.2 months) no patients had more than mild stenosis and 20 patients had mild or less regurgitation. None required aortic valve re-operation.
Conclusions: The Ozaki procedure has excellent short-term results and could be considered for aortic valve reconstruction in pediatric patients with congenital aortic valve disease. Longer-tem follow-up is necessary to determine the optimal patch material, valve function and

O2895 - TETRALOGY OF FALLOT INCIDENCE AND TIMING OF PULMONARY VALVE REPLACEMENT AFTER INITIAL REPAIR

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Background: The timing and incidence of pulmonary valve insertion after initial repair of tetralogy of Fallot (TOF) is important for strategic planning and counseling of families and may be influenced by initial surgical technique. We conducted a retrospective chart review of patients who underwent primary repair of TOF at our institution and assessed the timing and incidence of need for pulmonary valve replacement (PVR) after different surgical strategies.
Material and Methods: The surgical records of all patients treated at our institution for TOF from 1990 to 2016 were obtained. Patients were divided into 3 groups based on repair technique: 1) transatrial-transpulmonary, 2) transatrial-transpulmonary with

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infundibular patch, and 3) transannular patch. Patients were then evaluated for the incidence and timing of PV insertion.
Results: There were 327 patients who underwent TOF repair. Of these 160 (49%) had transatrial-transpulmonary repair, 89 (27%) had transatrial-transpulmonary repair with an infundibular patch, and 78 (24%) had transannular patch repair. A total of 76 (23%) prior shunt operations were performed in 7% (11/160), 25% (22/89), and 55% (43/78) of patients, respectively ($p < 0.001$). Median age at repair was 0.49 years. Nineteen patients (5.8%) underwent PVR following TOF repair (mean age was 13.7 ± 5.2 years). Of these 19 patients, 4 had a prior transatrial-transpulmonary approach, 2 had a transatrial-transpulmonary patch, and 13 had a transannular patch procedure ($p < 0.001$).
Conclusions: The placement of an infundibular patch did not increase the incidence of need for late PV insertion. Patients with a transannular patch were at the highest risk for undergoing a late PV insertion. Patients requiring a preliminary shunt operation have a much higher incidence of need for transannular patch at the time of repair for TOF. Our strategy of emphasizing valve preservation has resulted in only 6% of TOF repair patients undergoing late pulmonary valve insertion.

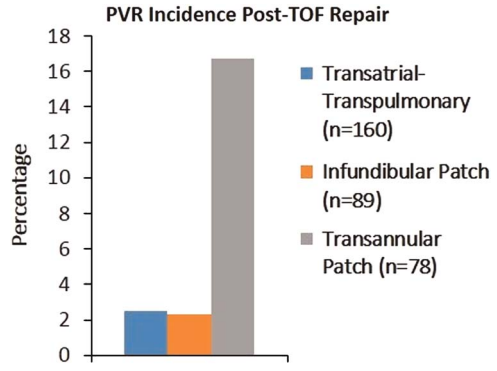

Figure.

O2929 - SURGERY ON DILATED AORTA ASSOCIATED WITH CONNECTIVE TISSUE DISEASE OR INFLAMMATORY VASCULITIS IN CHILDREN AND ADOLESCENT PATIENTS

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Background: We reviewed the patients who underwent surgery for dilated aorta, especially which was associated with connective tissue diseases or inflammatory vasculitis in children and adolescent.
Methods: Medical records of the patients who underwent aortic surgery for dilatation due to connective tissue diseases or inflammatory vasculitis between 2000 and 2016 were retrospectively reviewed. We excluded the cases of post-stenotic dilatation, acquired dilative aortic diseases, or secondary dilative change due to other congenital heart diseases or procedures.
Results: We have 12 patients who underwent surgery for dilated aorta. The median age was 10.2 years (5.4 months ~ 20.7 years), and the median body weight was 28.1 kg (6.8 kg ~ 81.5 kg). Associated diseases were Marfan syndrome (N = 4), Loeys-Dietz syndrome (N = 3), Kawasaki disease (N = 1), Takayasu's arteritis (N = 1), PHACE syndrome (N = 1), and Tuberous sclerosis (N = 1). Remaining 1 patient was supicious of having a connective tissue disease, however, not definitely confirmed. Most commonly involved area were aortic root and ascending aorta, but distal aortic arch was involved in 2 patient (unknown disease and tuberous sclerosis patients). Various surgical techniques were used to reduce dimension of dilated aorta and aortic root or aortic valve regurgitation, from simple aneurysmorrhaphy to David operation. For 79.2 ± 29.4 months of follow-up duration, there were 2 cases of follow-up loss in Marfan syndrome patients, and 5 reoperations in 3 patients. Except 1 patient, functional class and ventricular function were well maintained in all followed-up patients, however, the dimension of aortic root and involved aorta showed progressive dilation by time.
Conclusion: The cases of surgical treatment for the dilated aortopathy associated with connective tissue diseases and inflammatory vasculitis were very rare in children and adolescent. Most of the patients showed tolerable postoperative course, however, they showed progressive dilation of involved aorta by time even after surgical treatment. Close and regular follow-up is mandatory.

O3025 - IS THERE ANY ADVERSE EFFECT AFTER SYSTEMIC –TO PULMONARY ARTERY SHUNT IN PATIENTS WITH BIVENTRICULAR CIRCULATION 24 YEARS' EXPERIENCE AT OKAYAMA UNIVERSITY

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Objective: Recent trend of surgical management in complex congenital heart defects is one stage total repair rather than staged palliation. The main reason is the high mortality and morbidity after shunt. From database of STS and EACTS, the mortality after shunt is more than 5% even in patients with biventricular physiology. We report our 24 years' single institution experience of systemic –to-pulmonary artery shunts in patients with biventricular circulation.
Methods: From January 1993 to June 2016, 228 consecutive patients underwent Blalock-Taussig (BT) shunt or central shunt. Morphology included: tetralogy of Fallot (39), pulmonary atresia with ventricular septal defects (VSD) (55), with major aorto-pulmonary collaterals (MAPCAs) (22), pulmonary atresia with intact ventricular defect (20), double outlet right ventricle (23), transposition of the great arteries(TGA) with VSD and pulmonary stenosis (10), congenitally corrected TGA (7) and others (52). Median age was 64 days and median body weight was 4.6 kg (0.9-14 kg). Modified BT shunt was used to 226, original BT in 1 and central shunt in 1 patient. Surgical approach was by thoracotomy in 150 patients and median sternotomy in 78 patients. Most of the shunt in neonates and small infants was done by senior surgeons.
Results: No early mortality and morbidity. There was no in hospital re-intervention or re operation. Before total correction, 1 patient died 4 years after shunt (patient was lost to follow up after shunt). Shunt failure was observed in 1 patient with thrombosis in the shunt due to staphylococcus septicemia 52 days after the shunt.
Conclusions: There was no increased mortality and morbidity after the shunt in patients with biventricular circulation. Therefore, surgical management should be individualized to the patient.

TRANSPLANT/HEART FAILURE/PULMONARY HYPERTENSION

O1262 - MITOCHONDRIAL OXIDANT STRESS AND IRON DYSREGULATION IN DOXORUBICIN INDUCED CARDIOTOXICITY

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Background/Hypothesis: Doxorubicin (DOX) is an anthracycline-based chemotherapeutic agent used to treat a variety of cancers in both adults and children. The most serious adverse effect of DOX is a cumulative, potentially life-threatening cardiotoxicity associated with myocardial oxidative stress. Recent data suggest that DOX-induced oxidant stress may result from mitochondrial iron dysregulation. The aim of this study was to evaluate the impact of DOX on mitochondrial oxidant stress and iron handling in isolated neonatal cardiomyocytes.
Materials and Methods: Cultured neonatal rat cardiomyocytes (H9c2) were treated with 5 μM DOX ± 1 μM mito-tempo (MT), a specific mitochondrial targeted antioxidant, in complete media for 18 hours (time to maximum apoptotic response based on previous experiments). Control H9c2 cells were cultured in complete media without DOX. Oxidant stress was assessed by measuring thiobarbituric acid reactive substances (TBARS) reflecting lipid peroxidation. Mitochondrial iron transport was assessed by measuring changes in mRNA expression of the mitochondrial specific ATP-binding cassette sub-family B (ABCB) iron transporters using qPCR.
Results: Compared with untreated control cardiomyocytes, oxidant stress was increased 3-fold, and ABCB7 and ABCB8 expression was reduced by greater than 50% in response to DOX. MT treatment attenuated oxidative stress, and improved both ABCB7 and ABCB8 expression during DOX exposure (Table).
Conclusion: DOX treatment increases mitochondrial oxidant stress associated with decreased expression of two important mitochondrial ATP-binding iron transporters. A mitochondrial specific antioxidant partially restores both ABCB7 and ABCB8 expression and attenuates mitochondrial oxidant stress in response to DOX treatment. These results confirm that DOX-induced cardiotoxicity is associated with abnormal mitochondrial iron handling and oxidant stress. Mitochondrial ATP-binding iron transporters are novel potential therapeutic targets for preventing cardiotoxicity in response to DOX treatment.

Table.

	Control	DOX	DOX + MT
Oxidant Stress	1.00 ± 0.08	3.03 ± 0.13	1.48 ± 0.24
ABCB7 Expression	1.13 ± 0.26	0.45 ± 0.20	0.78 ± 0.26
ABCB8 Expression	1.37 ± 0.48	0.47 ± 0.15	0.74 ± 0.18

*ALL number reported as fold of control

O1287 - A REVERSIBLE AND IRREVERSIBLE RESPONSE TO HAEMODYNAMIC UNLOADING IN FLOW INDUCED PULMONARY ARTERIAL HYPERTENSION. IDENTIFICATION OF THE POINT OF NO RETURN IN PULMONARY VASCULAR DISEASE

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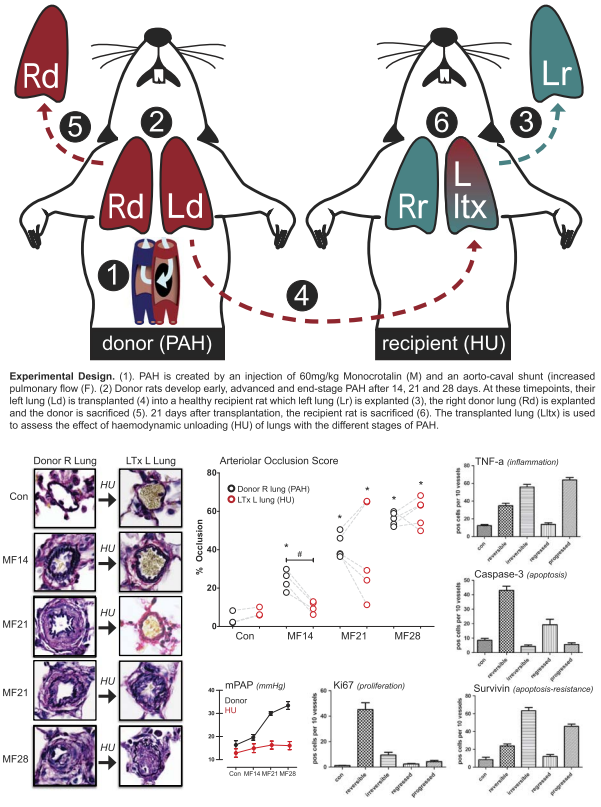
Background: Pulmonary arterial hypertension (PAH) is a progressive pulmonary vascular disease (PVD) that remains without a cure. In PAH associated with congenital cardiac shunts (PAH-CHD), timely haemodynamic unloading (HU) of the lungs by shunt closure may reverse this PVD, but not after a certain point of no return. The potential for reversibility in PAH has been established for decades, but mechanisms, timing and identification of such reversibility are still obscure today. We developed a novel rat model for HU in flow-induced PAH to study these mechanisms and to identify factors that specifically orchestrate the transition of reversible to irreversible PVD.
Methods: Flow-induced PAH was created in Lewis rats by injection of 60 mg/kg monocrotalin (t = 0) followed by a surgical aorto-caval shunt (t = 7days). To simulate HU, their left lung was transplanted into a healthy recipient rat at different stages of PAH development: early, advanced and end-stage PAH (t = 14/21/28). Recipient rats were sacrificed 21 days after HU.
Results: PAH developed time-dependently in donor rats, characterized by increased pulmonary artery pressure, vascular occlusion score and formation of neointimal lesions. HU normalized vascular occlusion ($p < 0.05$) in early-stage PAH by reducing medial hypertrophy: reversible PVD. In end-stage PAH, neointimal lesions did not reverse and arteries progressively occluded despite HU: irreversible PVD. In advanced PAH, the PVD was reversible in 3 and irreversible in 2 rats. The vascular phenotype



Experimental Design. (1). PAH is created by an injection of 60mg/kg Monocrotalin (M) and an aorto-caval shunt (increased pulmonary flow (F). (2) Donor rats develop early, advanced and end-stage PAH after 14, 21 and 28 days. At these timepoints, their left lung (Ld) is transplanted (4) into a healthy recipient rat which left lung (Lr) is explanted (3), the right donor lung (Rd) is explanted and the donor is sacrificed (5). 21 days after transplantation, the recipient rat is sacrificed (6). The transplanted lung (Lltx) is used to assess the effect of haemodynamic unloading (HU) of lungs with the different stages of PAH.

Figure.

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was more proliferative and pro-apoptotic in reversible PVD, and more inflammatory and apoptosis-resistant in irreversible PVD ($p < 0.0001$).
Conclusion: We have developed a rat model for flow-induced PAH with a reversible and irreversible response to HU, similar to the observation in children with PAH-CHD. This novel model offers opportunities to study pathophysiological and molecular mechanisms that orchestrate the transition from reversible to irreversible PAH. Identification of these mechanisms may provide new targets for pharmacological reversal of "irreversible" PAH.

O1308 - MACITENTAN VERSUS STANDARD OF CARE IN DELAYING DISEASE PROGRESSION IN CHILDREN WITH PULMONARY ARTERIAL HYPERTENSION THE TOMORROW STUDY DESIGN

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Background/Hypothesis: Management of pulmonary arterial hypertension (PAH) in children is primarily based on adult studies and short-term, non-randomized pediatric studies. The TOMORROW study was designed using information from large-scale pediatric PAH registries to evaluate macitentan versus standard of care (SoC) on clinically relevant long-term outcomes in pediatric PAH.
Materials and Methods: In this ongoing, prospective, long-term, randomized, open-label, phase 3 study (NCT02932410), patients aged 2-17 years with a PAH diagnosis ≤12 months before randomization and in WHO functional class (FC) I-III are randomized 1:1 to receive macitentan (concomitant phosphodiesterase type-5 inhibitor allowed) or continue SoC (according to investigator s judgment; ≤2 PAH-specific medications, including other endothelin receptor antagonists; excluding macitentan and intravenous/subcutaneous prostanoids). Macitentan dispersible tablet is administered orally once-daily, dosed based on body weight (3.5-10.0 mg/day). The composite primary endpoint is time to first disease progression event until end of study. Disease progression events: all-cause death; atrial septostomy, Potts anastomosis or lung transplant registration; hospitalization due to worsening PAH; clinical worsening of PAH (defined as need for/initiation of new PAH therapy or intravenous diuretics or continuous oxygen use, and ≥1 of the following: 1) worsening WHO FC, 2) new occurrence/worsening syncope, 3) new occurrence/worsening of ≥2 PAH symptoms [i.e. dyspnea, chest pain, cyanosis, near syncope, fatigue], or 4) new occurrence/worsening of right heart failure not responding to oral diuretics). Primary endpoint events are adjudicated by an independent Clinical Event Committee. Safety, tolerability and pharmacokinetics of macitentan are assessed.
Results: TOMORROW is powered based on the treatment effect observed in adults. To obtain 187 events, approximately 300 patients are planned for enrollment from >30 countries.
Conclusion: The TOMORROW study represents an advancement in the field of pediatric PAH. It is the first event-driven study among children with PAH and will assess long-term outcomes with a composite primary endpoint of disease progression.

O1354 - PROGRESS IN THE GENERATION OF MULTIPLE LINEAGE HUMAN IPSC DERIVED 3D ENGINEERED CARDIAC TISSUES FOR CARDIAC REPAIR.

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Despite improvements over the past 30 years, the 10-year pediatric heart transplantation survival rate remains low at 63%. In contrast to isolated cell therapies, implantable engineered cardiac tissues (ECTs) recover myocardial mass and function, creating the opportunity for recovery rather than replacement. Our ECT research has progressed from embryonic avian and rodent cell compositions to h-iPSC multiple cell lineage formulations with the goal of clinical translation. We generated ECTs from h-iPSC derived cardiomyocytes, endothelial cells, and vascular mural cells in both linear (15×1 mm) and large format (LF, 20×20 mm) geometries and are investigating the replacement of rodent matrix with human reagents. h-iPSC ECTs underwent rapid gel compaction and intrinsic beating initiated day3. CM fraction was similar at day14 for linear, LF, and all-human reagent ECTs (56 ± 8 vs. $57 \pm 21\%$ vs. $43 \pm 13\%$). ECT maximum capture rates were similar (4.5 ± 0.6 vs. 4.5 ± 1.1 vs 4.2 ± 1.2 Hz) as were active stresses (0.55 ± 25 vs. 0.54 ± 0.21 vs. 0.72 ± 0.18 mN/mm2). Prolonged culture to 28 days increased CM alignment concentration index for LF-ECT ($6.05 \pm 0.64^{\star}$ vs. 3.13 ± 0.33). Chronic optogenetic pacing did not change linear ECT CM alignment but did increased MCR at d14 (5.19 ± 0.37 Hz*) and ECT force-frequency relations shifted from negative to neutral. LF-ECTs implanted onto xenotolerant infarcted rat hearts improved ejection fraction (66.6 ± 5.8 vs. $49.3 \pm 13.2\%^{\star}$), normalized regional echo strain patterns, and reduced scar area (11.6 ± 7.6 vs. $23.9 \pm 7.2\%^{\star}$) at 4 weeks. ECT survival, engraftment, and perfusion were confirmed by histology. ($^{\star}P < 0.05$)
Conclusions: ECTs generated from h-iPSC using rodent and human matrix factors form functioning human myocardium that can survive in vivo implantation to recover cardiac structure and function. These compositions show promise as a strategy for pediatric myocardial recovery.

O1628 - CONTINUING CHALLENGES IN PEDIATRIC MECHANICAL CIRCULATORY SUPPORT ANALYSIS OF THE PEDIMACS REGISTRY

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Boston Children, Cardiology, Boston, Ma-United States[1]; Lucile Packard Children, Cardiology, Palo Alto, CA-United States[2]; Children, Cardiology, Philadelphia, PA-United States[3]; Cincinnati Children, Cardiac Surgery, Cincinnati, OH-United States[4] University of Alabama,

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Background: Expanded use of pediatric ventricular assist devices (VADs) has decreased mortality in children awaiting heart transplantation. PediMACS, an NIH-sponsored U.S. database, provides a unique platform to understand patient characteristics, device selection, and current needs. METHODS: Between 9/12–9/16, 42 US hospitals implanted 435 devices in 367 patients under 19 years old. Pre-implant characteristics, outcomes, and serious adverse events (SAE) (Infection, Bleeding, Neurologic, Device Malfunction) were analyzed.
Results: Diagnoses included cardiomyopathy in 224 (61%) pts and congenital heart disease in 78 (21%), of which 47 had single ventricle. Most (85%) were Intermacs level 1 or 2 at implant, with 152 (42%) intubated preimplant. Device type correlated with patient age. Median (IQR) age (yrs) at implant for paracorporeal continuous flow (PC) pumps (n = 60): 1.7 (0.3-10.0), for paracorporeal pulsatile (PP) pumps (n = 106):1.8 (0.4-5.4), and for implantable continuous flow (IC) pumps (n = 175): 15.0(11.2-16.8). Patients required LVAD in 295 (80%), BiVAD in 56 (15%) and total artificial heart in 8 (2%). Nearly 50% of patients underwent transplantation within 6 mos, with overall mortality before transplant of 18%. Actuarial survival on device at 6 mos. was 45% for PC, 64% PP, and 86% IC ($p < 0.0001$). Patients <5 years old had 60% 6 month survival, worse with CHD than CM (45% vs 68%, $p = 0.01$). At least one SAE occurred in 214 pts (58%), with a higher frequency in pts <5 years vs older (68% vs 53%, $p < 0.01$).
Conclusions: Pedimacs constitutes the largest longitudinal registry of pediatric VAD pts. Older children with continuous flow pumps have excellent outcomes. Smaller children, especially those with CHD continue to challenge caregivers, with higher incidence of SAEs and lower survival. Device development for small children remains a pressing need.

O1841 - EFFECT OF MIR 29B ON THE PROLIFERATION AND APOPTOSIS OF PULMONARY ARTERY SMOOTH MUSCLE CELLS BY TARGETING CCND2 AND MCL 1

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The proliferation and apoptosis of pulmonary artery smooth muscle cells (PASMCs) are considered to be key steps in the progression of pulmonary arterial hypertension (PAH). microRNAs (miRNAs) have been revealed as critical modulators of various diseases in which miR-29b is known to regulate cell growth and apoptosis by targeting CCND2 and Mcl-1. However, roles of miR-29b in PAH remain almost unknown. We hypothesized that miR-29b may control the proliferation and apoptosis progress by inhibiting its own specific targets in PASMCs. miR-29b significantly suppressed the proliferation and promoted apoptosis of PASMCs through the inhibition of CCND2 and Mcl-1.We also found that miR-29b expression levels significantly declined in pulmonary artery tissue of PAH model induced by monocrotaline (MCT) in rat and hypoxia in mouse and that the overexpression of miR-29b can inhibit the expression of CCND2 and Mcl-1 and reduce the right ventricular systolic pressure (RVSP) and right ventricular hypertrophy (RVHI). Consistent with the important role of miR-29b in vitro, we observed down-regulated expression levels of CCND2 and Mcl-1 from the PAH model induced by MCT in rat. These results demonstrated that miR-29b suppressed the proliferation and promoted apoptosis of PASMCs, possibly through the inhibition of CCND2 and Mcl-1, and suggest that miR-29b may serve as a useful therapeutic tool to treat PAH.

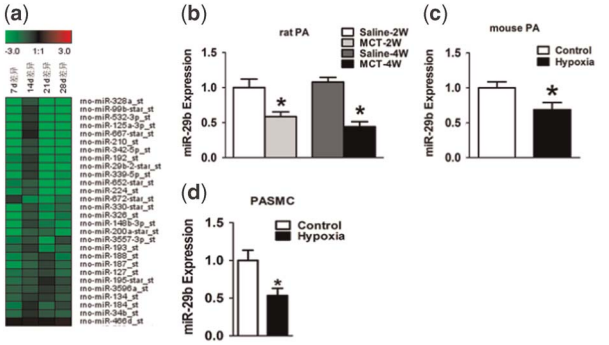

Figure 1.
*miR-29b is down regulated in PAH. (a) Partially differential expression of miRNAs in model of PAH induced by MCT for 7days, 14days, 21days, 28days, Cluster analysis of miRNAs expression from individual specimens were assessed by microarray analysis. (b) miR-29b expression in pulmonary artery tissue of PAH model induced by MCT in rat by real-time PCR assay. (c) miR-29b expression in pulmonary artery tissue of PAH model induced by hypoxia in mouse. (d) miR-29b expression in PASMCs after hypoxic treatment 48h (*p < 0.05).*

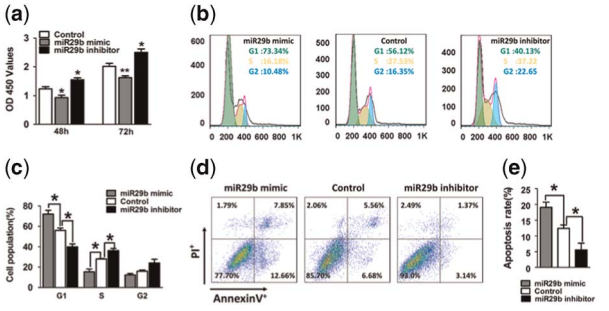

Figure 2.
*miR-29b affects the proliferation and apoptosis of PASMCs. (a) OD of PASMCs after transfection with miR-29b mimic or inhibitor at different cultured times, CCK8 assays revealed that upregulation of miR-29b inhibited the number of PASMCs. (b,c) Cell cycle analysis of PASMCs treated with miR-29b mimic, inhibitor or control and cultured for 48 h after cell transfection. (d,e) Apoptosis rate of PASMCs treated with miR-29b mimic or inhibitor and cultured for 48 h after cell transfection (*p < 0.05, ** p < 0.01).*

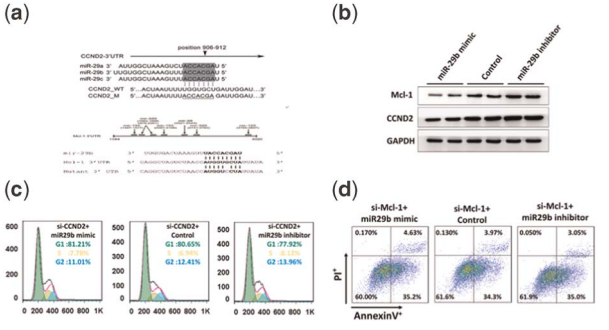

Figure 3.
miR-29b targets 3'UTR of CCND2 and Mcl-1 and inhibits expression of CCND2 and Mcl-1. (a) 3'-UTR of CCND2 and Mcl-1 binding sites in miR-29b. (b) Western blot analysis of expression of CCND2 and MCL-1 protein in PASMCs treated with miR-29b mimic, inhibitor, or control. GAPDH served as the loading control. (c) Effect of si-CCND2 silencing CCND2 expression on PASMCs cell cycle. (d) Effect of si-Mcl-1 silencing Mcl-1 expression on PASMCs apoptosis rate.

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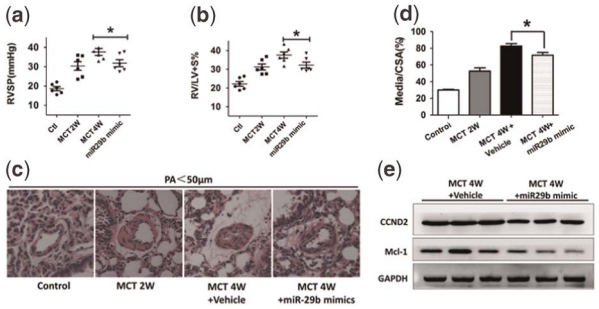

Figure 4.
*miR-29b attenuates MCT-induced pulmonary vascular remodeling. Right ventricular systolic pressure (RVSP) (a) and right ventricular hypertrophy [RV/ (LV+ S)] (b) are shown in bar charts. Data are shown as means ± SE (n = 6). (c,d) The wall thickness of small and medium sized pulmonary arteries was assessed from HE stained lung sections. Bar charts showing the percentage of wall thickness to vessel external diameter of each rat group. Data are shown as means ± SE (n = 6). (e) Comparison of protein expression between CCND2 and Mcl-1 in the two groups (*p < 0.05).*

O1921 - ALTERED AORTIC VALVE LEAFLET GENE EXPRESSION FOLLOWING CONTINUOUS FLOW VAD THERAPY IN PEDIATRIC PATIENTS

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Introduction: Use of continuous flow LVAD for bridge to transplant therapy in pediatric heart failure patients is becoming a mainstay. It has been known that in these patients AV leaflets start to develop insufficiency, resulting in LVAD failure due to circulatory shunt. We sought a better understanding of the effects consequent to disrupted AV closure on leaflet tissue. Accordingly, we have used a transcriptome-wide gene expression analysis approach to compare patients with and without durable LVAD therapy.
Methods: We analyzed specimens from two groups of pediatric heart AV leaflets (AVL): Patients on durable continuous flow LVAD (LVAD, N = 4) and normal pediatric donor hearts (Con, N = 4). All specimens were normal in gross appearance. RNA was extracted, labeled and used to interrogate Affymetrix human whole transcriptome arrays.
Results: Genome-wide expression level comparison demonstrated that LVAD AVL exhibited more than 400 genes differentially regulated significantly more than Con AVL (>2.5 fold; P < 0.05).
Conclusions: We conclude that AVL undergo pathogenic changes during durable chronic LVAD therapy, manifest as alteration in gene expression. Our results therefore support the concept that altered AV cycling pattern during continuous flow LVAD therapy induces early stages of AVL pathogenesis. The data suggest that AVL homeostasis requires a normal pattern of frequent valve coaptation and opening that is abrogated by LVAD therapy.

O2035 - PILOT STUDY OF WARFARIN GENOTYPE POLYMORPHISMS IN CHILDREN WITH VENTRICULAR ASSIST DEVICES. (VAD)

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Hypothesis: The objective of our study was to determine the incidence of warfarin polymorphisms (VKORC1, CY2CP9 and CYP4F29) in children with VAD implantation and evaluate the correlation between bleeding and thrombo-embolic events. We analysed the genotype polymorphism profile in 24 children with VAD implantation.
Materials and Methods: Children who had undergone VAD implantation from the cardiac department of the Children's Hospital Bambino Gesu' were recruited. A combined retrospective, prospective study was performed. Charts were analyzed and we recorded: polymorphisms, disease, type of VAD implanted, race, age, gender, height, weight, bleeding and thrombo-embolic episodes. Blood samples were obtained during ambulatory visits. Informed consent was obtained.
Results: Mean weight was 19,96 kg (SD 20,69 kg), mean age was 68,18 months (SD 74 cm). Twenty one patients had a diagnosis of dilated cardiomyopathy and 3 patients with a restrictive cardiomyopathy. Five Jarvik 2000's were implanted, 18 Berlin Hearts and 1 patient with a Heartmate3. Seventy-two percent underwent heart transplantation. Sixteen percent of the patients bleed during VAD implantation and the site of bleeding was always cerebral. The incidence of thrombosis was 52%, 9 patients showed device thrombosis and 7 brain thrombosis. Twelve percent of patients had neurological damage pre-VAD, 36% percent had post_VAD neurological damage. Statistical analysis showed a partial correlation between thromboembolic events and the presence of the CYP4F2 polymorphism with a significant trend (p = 0.07). Moreover, the CYP2C9 showed a positive predictive value of 86% for thromboembolism. In addition, we found an incidence of 36% of the CYP4F2 polymorphism.
Conclusions: Our study underlines the usefulness and importance of analysing warfarin polymorphisms in this particular group of patients. We found a partial correlation between thromboembolic events and the CYP4F2 polymorphism which also had a higher incidence compared with the general population. Our results are preliminary and the group of patients was small.

O2039 - POTTS SHUNT RESULTS IMPROVES RIGHT VENTRICULAR FUNCTION IN PATIENTS WITH SUPRASYSTEMIC PULMONARY HYPERTENSION.

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Backgrounds and Objective: Pulmonary arterial hypertension (PAH) carries a poor prognosis. Right ventricular (RV) function and work are pivotal to survival in PAH. Recent application of left pulmonary artery to descending aorta shunt, Potts shunt, to off-load the RV has demonstrated promising clinical results. Studies to evaluate the impact of Potts shunt on RV mechanics that may impact survival are lacking. We sought to mechanistically evaluate RV function and work before and after surgical Potts shunt in pediatric PAH patients.
Method: In a retrospective study with prospective analysis, we assessed echocardiographic indices before and after (median 7 ± 8.2 days) a Potts shunt was performed in 10 consecutive pediatric patients (median age 11.8 ± 4.8 years, weight 35.4 ± 20.3 kg) with suprasystemic PAH. We measured tricuspid annular plane systolic excursion (TAPSE) to assess RV systolic function; tricuspid regurgitation (TR) jet velocity to estimate pulmonary artery systolic pressure (PASP);

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pulmonary artery acceleration time (PAAT) as well as its ratio to right ventricular ejection time (RVET) as indices of pulmonary artery pressure and resistance; and TAPSExPASP and TAPSExPAAT to develop length-force relationship as indices of RV work. We analyzed pre and post shunt data using Wilcoxon signed-ranked test. A $p < 0.05$ was considered significant.

Result: After the Potts shunt, RV systolic function (TAPSE % increased 17.5 ± 15.5) and pulmonary artery hemodynamics (PAAT % increased $38.4\% \pm 23.7$) significantly improved (both $p = 0.005$) but PASP did not ($p = 0.721$). RV work significantly improved (% increased of TAPSExPAAT 66.4 ± 34.9 and TAPSExPASP 17.5 ± 15.6; $p < 0.05$). (Figure 1)

Conclusions: RV function and RV work improve rapidly after the Potts shunt. This initial evidence of improved RV function and work may indicate a protective effect of the Potts shunt on the RV that may aid survival in pediatric patients with PAH.

	Pre	Post	Difference	P-Value
TAPSE	1.28±0.15	1.50±0.15	18.4%±15.9	0.005*
PASP	87.8±21.7	90.8±20.7	6.8%±23.2	0.721
PAAT	74.7±13.2	103.1±15.0	40.7%±23.7	0.005*
PAAT/RVET	0.381±0.108	0.441±0.045	22.0%±25.7	0.075
TAPSExPASP	113.5±35.4	135.8±31.4	19.6%±29.9	0.047*
TAPSExPAAT	95.6±19.4	155.5±31.8	66.4%±34.9	0.005*

Abbreviations: tricuspid annular plane systolic excursion (TAPSE), pulmonary artery systolic pressure (PASP), pulmonary artery acceleration time (PAAT), right ventricle ejection time (RVET).

Figure 1.
Right Ventricular Mechanics Before And After Potts Shunt.

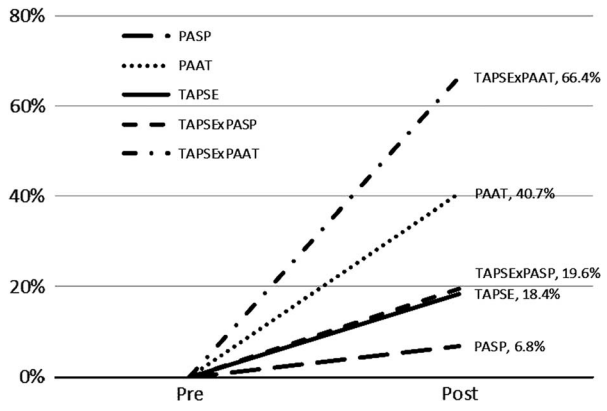

Figure 2.
Changes In RV Mechanics After Potts Shunt.
Abbreviations: right ventricule (RV), tricuspid annular plane systolic excursion (TAPSE), pulmonary artery systolic pressure (PASP), pulmonary artery acceleration time (PAAT), right ventricle ejection time (RVET).

O2081 - PROSPECTIVE ECHOCARDIOGRAPHIC EVALUATION OF VENTRICULAR LOADING TRENDS IN PEDIATRIC PATIENTS WITH PULSATILE FLOW LVAD

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The aim of this study was to describe the echocardiographic trend of LV and right ventricular (RV) function after implantation of a pulsatile flow LVAD in children.. From December 2013 to January 2017, we prospectively enrolled all consecutive pediatric LVAD (Berlin Heart Excor) patients collecting clinical and echocardiographic data at baseline, within 24 hours after implantation and monthly until LVAD explantation. Fourteen patients were enrolled with an average age and weight at the implantation of 12.4 ± 11.0 months and 7.0 ± 3.6 kg. Thirteen (93%) were affected by dilated cardiomyopathy and 1 by restrictive cardiomyopathy. The average LVAD stay was 226.2 ± 121.2 days. Ten (71%) were successfully transplanted, 4 (30%) died (3 because of major neurological complications and 1 after LVAD explantation used as a bridge to recovery). After an average LVAD stay of 120 days, 4 patients required prolonged inotropic support for RV failure. LV end-systolic and end-diastolic volumes were reduced in the acute phase ($p = 0.034$ and $p = 0.004$), at one month ($p = 0.0075$ and $p = 0.0005$) and two months ($p = 0.019$ and $p = 0.001$). RV function improved in the acute phase, but then it started to decrease over time with a progressive increase in RV dimensions. After 4 months follow up, RV fractional area change (RVFAC) worsening was related with the deterioration of LV unloading ($p = 0.0036$) with need of prolonged inotropic ($p = 0.032$). Pulsatile LVAD in pediatrics is followed by an early and mid-term LV reverse remodeling due to a change in loading conditions, yet at long term follow it does not remain stable. RV function improved in the acute phase, but a progressive RV dilatation was noted leading to the need of prolonged inotropic. The potential of heart recovery seems to be more promising during the early phase post LVAD. Further studies are needed to understand how to prolong the benefits provided by the LVAD.

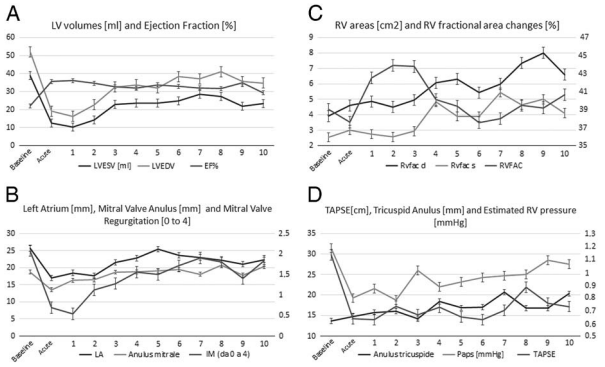

Figure.

O2336 - NEONATAL EBSTEIN'S ANOMALY WHAT'S TO EXPECT

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Utrecht, Wilhelmina Children's Hospital, Pediatric Cardiology, Utrecht-The Netherlands[8]

Background: Ebstein's anomaly is a rare disease with a wide clinical spectrum. Forecasting the prognosis of neonates with Ebstein's anomaly is difficult and only few outcome studies are available.
Materials and Methods: In the Netherlands, all consecutive live-born patients diagnosed with Ebstein's anomaly in the neonatal period (0-30 days) between 1980 and 2014 were included. Survival curves were obtained using the Kaplan–Meier method. By using the Cox proportional hazard model, we analyzed factors associated with death.
Results: We included 112 neonates (51 male) of whom 65 (58%) presented in NYHA Class IV. Twenty-four (21%) showed severe right ventricular outflow tract obstruction, in 34 (31%) the tricuspid regurgitation was severe. The mean follow-up was 85 months (0-216 months). Ten patients underwent a percutaneous intervention (four balloon pulmonary valvuloplasty procedures, three Rashkind procedures, two radio-frequency perforations of the pulmonary valve and one radio-frequency ablation because of refractory supraventricular tachycardia). Neonatal surgery was required in 21 patients (19%), five required one or more re-operations within the first year of life. In total, twenty-seven patients (24%) died; 16 within the neonatal period and all but two died within the first year of life. The majority (12 patients) died of heart failure. The one-month survival was 86%, the one-year-survival 77%. Factors associated with death included presentation in NYHA Class IV (HR 10.7, 95% CI 2.5-45,1, $p = 0.001$), severe right ventricular outflow tract obstruction (HR 2.5, 95% CI 1.1-5.4, $p = 0.024$) and neonatal surgery (HR 9.6, 95% CI 1.2-78.7, $p = 0.034$).
Conclusion: We present the results of one of Europe's largest cohorts of neonates with Ebstein's anomaly. Twenty-four percent died, all but two during the first year of life. Presentation in NYHA Class IV, severe right ventricular outflow tract obstruction and neonatal surgery were associated with death.

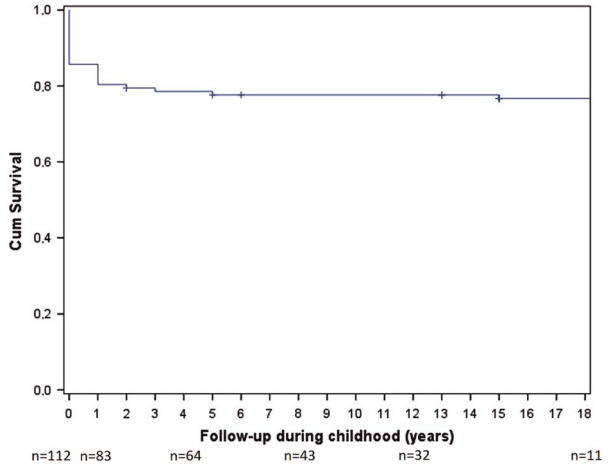

Figure 1.
Survival during childhood and adolescence (0-18 years) of 112 neonates with Ebstein's anomaly.

O2391 - SHORT TERM PROGNOSTIC INDICATORS IN ADULT CONGENITAL HEART DISEASE PATIENTS WITH HEART FAILURE

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Background: Although heart failure (HF) is the leading cause of premature death in adult congenital heart disease (ACHD), little population-specific data exist.
Objectives: This study reports early experience from a dedicated, sub-specialty adult congenital heart disease-heart failure (ACHD-HF) clinic to identify risk factors for adverse outcome.
Methods: Between 2012 and 2015, 126 patients (57% male) attended the ACHD-HF clinic. Baseline and follow-up data were analyzed and compared across 4 anatomical/physiological subgroups: cyanotic ACHD, single ventricle (1 V), biventricular circulation-subaortic right ventricle (2V-RV) and biventricular circulation-subaortic left ventricle (2V-LV). Multivariable Cox proportional hazard models were used to identify predictors of the composite primary outcome: death, transplant or ventricular assist device (VAD).
Results: Mean age at first visit was 38 ± 13 years. Patients were grouped as follows: cyanotic ACHD 10%, 1 V 24%, 2V-RV 29% and 2V-LV 37%. During a median follow-up of 1.7 (interquartile range: 0.8-2.9) years, 38 patients (30%) reached the primary outcome with event-free survival 89%, 78% and 63% at 1, 2 and 3 years respectively. Forty (31.7%) patients experienced 69 HF hospitalisations. Between-group differences were noted for systolic function, valvular regurgitation, pacing prevalence and invasive hemodynamics. Multivariable analysis revealed 2V-RV subgroup ($p = 0.001$), NYHA class ($p = 0.002$) B-type natriuretic peptide >164 pg/ml ($p = 0.003$) and sodium <136 mmol/L ($p = 0.036$) were independently associated with death, transplant or VAD.
Conclusions: Our young ACHD-HF patients experienced high adverse event rates during a limited period of follow-up. The prognostic markers identified by this study will aid clinicians to stratify short-term risk and thereby guide advanced HF management decisions in ACHD.

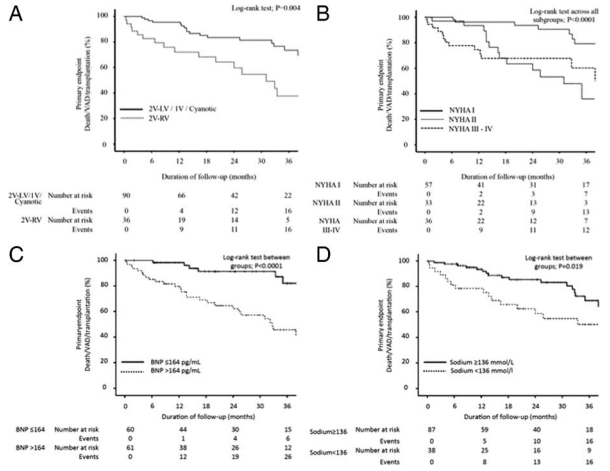

Figure.
Kaplan-Meier analyses of variables related with the primary endpoint (death/VAD/transplant) in multivariable Cox-regression analysis.

O2396 - REDUCED EXERCISE PERFORMANCE IS RELATED TO NUMBER OF THORACIC SURGERIES IN CHILDREN WITH CORRECTED TETRALOGY OF FALLOT OR FONTAN CIRCULATION

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Background: Adults with congenital heart defects (CHD) have a reduced exercise performance related to morbidity and mortality. It is unknown if this is due to changes in the circulation because of their CHD or due to other factors. Exercise performance and the relation with morbidity and mortality has been poorly described in children. The aim of this study was to describe exercise performance in children with CHD, and to describe the effect of thoracic surgery on exercise performance.
Materials and Methods: A retrospective study was performed between January 2012 and December 2016. Children with a corrected Tetralogy of Fallot (cTOF n = 75) or Fontan circulation (Fontan n = 39) between 8 and 18 years of age performed a cardiopulmonary exercise test (CPET) on a bicycle ergometer with a continuous RAMP-protocol. Increment protocols were based upon body height and estimated fitness. Exclusion criterion was respiratory expiratory ratio <1.00. Predictive values were calculated separately for boys and girls based on age (Bongers 2012). The total number of thoracic surgeries per individual were analyzed.
Results: Data is presented in a supplemental table. Both groups were limited in their exercise performance (predicted VO2peak cTOF 81 ± 22% and Fontan 67 ± 19%), Fontan significantly worse than cTOF (p = 0.002). One-way ANOVA showed significant lower exercise performance between the surgical groups (VO2% predicted p <0.001). Univariate linear regression showed a significant effect of number of thoracic surgeries on VO2%predicted (B -7.2%, p <0.001). In a multivariate model including diagnosis (cTOF and Fontan) and body weight, three or more thoracic surgeries showed significant effect of -11.6% (p = 0.021).
Conclusion: We showed that exercise performance is reduced already in young children with CHD. Children with Fontan are more impaired than those with cTOF. Independent of diagnosis and body weight, three or more thoracic surgeries had a negative effect on exercise performance.

Table.

	cTOF (n = 75)	Fontan (n = 39)	*p*-value
Patient characteristics			
Age (years)	137 ± 2.7	13.1 ± 2.9	0.276
Body length (cm)	160 ± 14	155 ± 16	0.154
Body weight(kg)	50 ± 14	46 ± 16	0.277
BMI (m^2/ke)	19.1 ± 3.7	18.5 ± 3.5	0.430
BSA (m^3)	1.47 ± 0.27	1.40 ± 0.32	0.217
Surgical data	n = 72	n = 38	
Total number of thoracotomies	119	119	
Lateral thoracotomies	28 (24%)	29 (24%)	
Medial sternotomies	94 (79%)	90 (76%)	
Age correction (years)	1.02 (0.84-1.25)	5.49 (3.78-6.70)⋆	<0.001
Exercise performance			
Workload (W)	143 ± 44	117 ± 40⋆	0.002
Workload Index (W/kg)	2.9 ± 0.6	2.6 ± 0.4⋆	0.001
Workload % predicted	81 ± 20	70 ± 15⋆	0.004
VO2peak index (ml/min/kg)	36.7 ± 7.3	31.0 ± 6.9⋆	<0.001
VO2peak% predicted	81 ± 22	67 ± 19⋆	0.002
Max. heart rate (BPM)	179 ± 14	166 ± 19⋆	<0.001
Heart rate reserve (BPM)	21 ± 14	34 ± 19⋆	<0.001
Minute ventilation index (L/min/ke)	1.21 ± 0.29	1.17 ± 0.30	0.408
Minute ventilation % predicted	77 ± 20 n = 73	75 ± 20 n = 14	0.511
VE/VCO2 slope	25.6 ± 4.0	28.6 ± 4.2⋆	0.011

Supplemental table. BMI body mass index. BSA body surface area. In case of a normal distribution variables are shown as mean ± SD and tested with an Independent T-test ($p = 0.05$). In case of non-parametric distribution Variables are shown as median (IQR) and tested with a Mann-Whitney U test ($p = 0.05$).
⋆ <0.05 Fontan vs. cTOF

O2398 - OUTCOME FOLLOWING ASSESSMENT OF PAEDIATRIC PATIENTS FOR CARDIAC TRANSPLANT

Rebecca Johnson[1], Louise Coats[1], Kate Best[2], Gareth Parry[3], Zdenka Reinhardt[1], John Osullivan[1], Emma Simpson[4], Fabrizio De Rita[5], Asif Hasan[5], David Crossland[1]
Freeman Hospital, Congenital Cardiology, Newcastle Upon Tyne-United Kingdom[1]; Newcastle University, Institute of Health and Society, Newcastle Upon Tyne-United Kingdom[2]; Freeman Hospital, Department of Cardiothoracic Transplant, Newcastle Upon Tyne-United Kingdom[3]; Freeman Hospital, Paediatric Cardiac Intensive Care, Newcastle Upon Tyne-United Kingdom[4]; Freeman Hospital, Congenital Cardiac Surgery, Newcastle Upon Tyne-United Kingdom[5]

Background: Outcomes following paediatric cardiac transplant are well documented. Less is known about outcomes from the time of assessment and listing. We aimed to describe the outcomes of children assessed for cardiac transplant comparing those with congenital heart disease (CHD) and cardiomyopathy (CM).
Methods: A retrospective case note analysis of all children (0-18 years) assessed for cardiac transplant at Freeman Hospital, Newcastle upon Tyne, UK between 2005 and 2015 was performed.
Results: 256 patients were assessed (54.3% male, median age 2.5 IQR 0.7-9.8 years, 43.4% CHD, 56.6% CM). There was no difference in age between the two groups (CHD: 3.6 IQR 0.8-9.7 years v. CM: 2.0 IQR 0.7-10.4 years, p = 0.371) or the proportion listed for transplant (CHD:72% v. CHD:77%, p = 0.344). Mechanical support was used less frequently in the CHD group (CHD:21% v. CM:48%, p < 0.0001). Survival following assessment was worse in CHD patients (CHD – 1 year:65%, 5 year: 59% v. CM – 1 year:83%, 5 year:79%, p = 0.008) and a higher proportion of CHD patients died on the waiting list (CHD:29% v. CM:13%, p = 0.009). Of those listed, fewer CHD patients underwent transplant (CHD: 61% v. CM:78%, p = 0.014). Post-transplant survival was comparable between groups (CHD - 1 year:90%, 5 year:82% v. CM - 1 year:93%, 5 year:88%, p = 0.584).
Conclusions: Following assessment for cardiac transplant CHD patients have a higher mortality, they are more likely to die on the waiting list and are less likely to be transplanted than those with CM. This is despite similar rates of listing and equivalent post-transplant survival. This is partly explained by the limited suitability of CHD patients for mechanical support. These findings should be considered when determining urgent transplant listing criteria and priority for heart allocation in children and highlights the need for more effective mechanical support in the CHD population.

O2577 - OUTCOMES AFTER ORTHOTOPIC HEART TRANSPLANT AT TEXAS CHILDREN'S HOSPITAL FOCUS ON RETRANSPLANTATION

Rodrigo Zea-Vera[1], Nathan Rodgers[2], Huirong Zhu[3], William J. Dreyer[2], Susan W. Denfield[2], Jack F. Price[2], Antonio G. Cabrera[2], Hari Tunuguntla[2], Claire Bocchini[4], Carlos M. Mery[1], Lauren C. Kane[1], Jeffrey S. Heinle[1], Charles D. Fraser[1], Iki Adachi[1]
Texas Childrens Hospital, Congenital Heart Surgery, Houston-United States[1]; Texas Childrens Hospital, Pediatric Cardiology, Houston-United States[2]; Texas Childrens Hospital, Outcomes and Impact Center, Houston-United States[3]; Texas Childrens Hospital, Pediatrics, Houston-United States[4]

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Background: Controversies exist regarding repeat heart transplant due to its poorer outcome compared to primary transplant in the setting of organ shortage. We sought to review outcomes at our institution, to guide the patient selection process.
Materials and Methods: All patients listed for isolated heart transplant at our institution between 1995-2016 were included. Wait-list and post-transplant outcomes were compared between the two groups: listing for primary (P) and repeat transplant (R). Poor wait-list outcome was defined as either death or removal from the wait-list due to poor condition. Post-transplant graft loss was defined as death or need for re-transplant. Waitlist outcomes were assessed using competing risk analysis. Post-transplant outcomes were evaluated using log-rank test with logistic regression.
Results: In total, 380 patients were listed 413 times: 360 and 53 in Group P and R, respectively. Overall, there was a difference in the proportion of patients that reached transplant: 72% and 58%, respectively ($p < 0.05$). At 1 year after listing, patients in Group R were less likely to have received transplant (P, 66% [95% CI:64-69] and R, 44% [95% CI:37-50]) and more likely to remain on the wait-list (P, 18% [95% CI:16-20] and R, 35% [95% CI:29-42]). Post-transplant graft survival was significantly different: 10-year graft survivals were 60% and 36%, respectively ($p = 0.02$). This was despite identical 1-year survival (92% and 90%, respectively), suggesting late attrition being the fundamental problem. In Group P, there was a reduction in graft loss over time: 10-year graft survival before and after 2005 were 49% and 68%, respectively ($p < 0.01$). In Group R, however, such a positive era-effect was absent: 7-year graft survival were 52% and 45%, respectively ($p = 0.57$). Years from previous transplant was a protective factor (OR:0.7 [95% CI: 0.5-0.9]) from graft loss in Group R.
Conclusions: Patients undergoing repeat heart transplant carry poorer wait-list and post-transplant outcomes. Further studies are warranted for better patient selection.

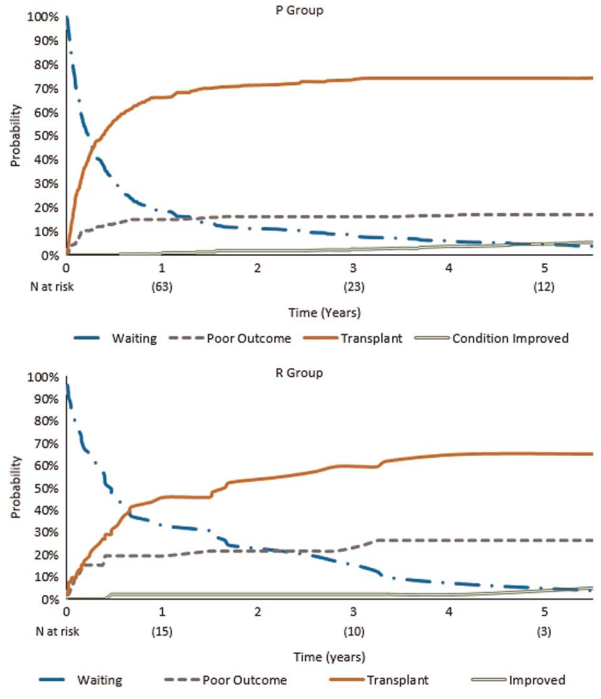

Figure.
Competing Risk Analysis for Waitlist Outcomes.

O2622 - EARLY RESULTS AND LONG TERM FOLLOW UP DOUBLE PATCH FLAP VALVE VSD CLOSURE IN PATIENTS WITH PULMONARY HYPERTENSION AND ELEVATED PULMONARY VASCULAR RESISTANCE.

William Novick[1], Vasily Lazorhyshynets[2], Alexander Govolenko[2], Frank Molloy[3], Marcelo Carderelli[4], Sara-Catherine Zzing[5], Thomas Di Sessa[6]
University of Tennessee Health Science Center, Surgery, Memphis-United States[1]; Amosov National Institute of Cardiovascular Surgery, Congenital Cardiac Surgery, Kyiv-Ukraine[2]; William Novick Global Cardiac Alliance, Intensive Care, Memphis-United States[3]; William Novick Global Cardiac Alliance, Surgery, Fairfax-United States[4]; University of Tennessee Health Science Center, College of Medicine, Memphis-United States[5]; William Novick Global Cardiac Alliance, Cardiology, Lexingotn-United States[6]

Background: Children with VSD, pulmonary hypertension (PHT) and elevated pulmonary vascular resistance (PVR) are at increased risk for both early and late death following VSD closure. We developed a fenestrated flap valve unidirectional VSD closure patch (DFV) to use in these patients. Our first implant was in 1996 and late follow-up (f/u) has been conducted on all patients at the initial institution.
Methods: Records from the institute where the first implant was performed (5/96) and from the NGO were queried for DFV patients. Beginning in 2005 all patients considered for DFV we placed on 3 months of sildenafil before catheterization. Initial and f/u records were pulled and analysis was performed on demographics, catheterizations, echocardiographic follow-up and mortality.
Results: Forty (40) patients received the DFV, hospital discharge survival was 97.5% (39/40). Twenty-two were female. Median age was 7.5 years. Baseline catheterization data: mean PAp 69.2 ± 9.6, median PVR 9 Wu, median PVR/SVR 0.50 and Qp/Qs 1.3. Pre-cath sildenafil did not significantly change PVR, Qp/Qs, or percentage of patients who responded to vasoactive testing (VAT), $p = 0.5$. Two patients were lost to follow-up at 5 and 5.5 years, there was 1 late death. The median f/u was 15.9 years. Risk factors for development of severe PHT at last f/u by univariant analysis were PVR/SVR of $\geq$ 0.56 ($p < 0.001$) pre-op, PVR >13.9 Wood units ($p < 0.01$) and a less than 10% change with VAT (oxygen only). The degree of PHT at last f/u in all patients was: none 8/38 (21.1%); mild 15/38 (39.5%); moderate 8 (21.1%) and severe 7/38 (18.4%).
Inferences: Young patients with PHT and severely elevated PVR have a significant chance of not developing severe PHT and therefore should not be denied operation routinely. An individualized approach is warranted.

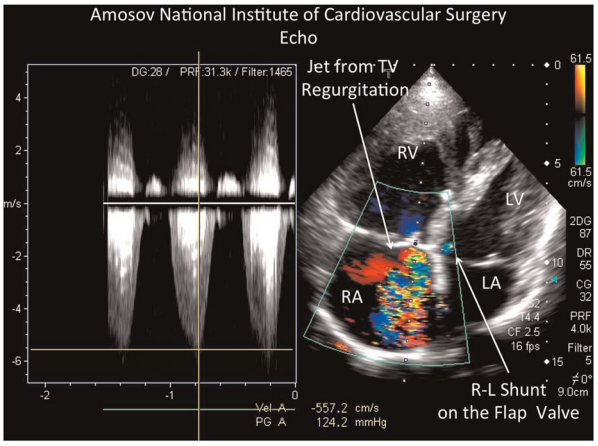

Figure.

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O2778 - RIGHT ATRIAL FUNCTION IN CHILDREN WITH PULMONARY HYPERTENSION.

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Stollery Childrens Hospital,University of Alberta, Pediatric Cardiac Critical Care and Pulmonary Hypertension, Edmonton-Canada[1]; Stollery Childrens Hospital,University of Alberta, Pediatric Cardiac Critical Care and Pulmonary Hypertension Service, Edmonton-Canada[2]; Stollery Childrens Hospital, University of Alberta, Pediatric Cardiac Critical Care Unit, Edmonton-Canada[3]; Stollery Childrens Hospital University of Alberta,, Pediatic Cardiology, Edmonton-Canada[4]; Stollery Childrens Hospital,University of Alberta, Pediatric Cardiology, Edmonton-Canada[5]; University of Alberta, Electrical and Computing Engineering, Edmonton-Canada[6]; Stollery Childrens Hospital, University of Alberta, Pediatric Pulmonary Hypertension, Edmonton-Canada[7]

Background: We sought to investigate whether right atrial (RA) area change, a marker of RV diastolic function, influences survival in childhood PAH.
Methods: We studied RA function in children with PAH undergoing cardiac catheterization and an echocardiogram between 2009-16. RA and RV endocardial area were traced in the apical 4 chamber view. We recorded maximum RA area, RA area at the onset of the P wave (RA area p) and minimum RA area. We calculated the following: RA(RAFAC) and RV (RVFAC) % fractional area change from (maximum RA or RV area - minimum RA or RV area)/ maximum RA or RV area), RA passive emptying (RAEp) from (maximum RA area – RA area p)/ maximum RA area, active RA emptying (RAEa) from (RA area p - minimum RA area)/ RA area p, RA active emptying fraction % (RAEF) from (RAFAC – RAEp)/RAFAC. Event free survival was the time from catheterization to starting parenteral treprostinil therapy, transplant or death.
Results: We studied 57 children (27 F), median age 3 yrs (range 0.3-17), BSA 0.56 m2 (0.2-1.8). Median event free survival was 3yrs (0.2–8.4),RAFAC was 34% (13-74), RAaEF was 60% (13-94%). Patients with RAaEF $\geq$ 58% had greater mean PA pressure 57 ± 22 mmHg vs. 38 ± 11 mmHg ($p = 0.0002$), PVRI 14.5 ± 7.7 vs. 8 ± 4 WU.m2 ($p = 0.0003$). RAaEF $\geq$ 58% was associated with decreased 5 yr event free survival (sensitivity of 100%, specificity of 56%). RVFAC $< 25\%$ was associated with worse survival at 5 yrs with a sensitivity of 72% and specificity of 79%. RAaEF $\geq$ 58% with RVFAC $< 25\%$ were independently associated with event free survival at 5 yrs ($p = 0.02$ and $p = 0.0003$ respectively).
Conclusion: In children with PAH impaired RV function is associated with poor outcomes. Increased RAaEF predicts decreased event free survival independently of RV systolic dysfunction.

O2823 - HEART RATE REDUCTION STRATEGY IN DUCHENNE DILATED CARDIOMYOPATHY

Rachele Adorisio[1], Nicoletta Cantarutti[1], Adele D[2], Anwar Baban[1], Fiore Iorio[1], Antonio Amodeo[1], Fabrizio Drago[1]
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Background: Dilated cardiomyopathy (DCM) is a major source of morbidity and mortality in Duchenne muscular dystrophy (DMD) patients. No studies are available on the effect of heart rate reduction (HRR) on DCM-DMD.
Objective: To evaluate the effectiveness of a HRR strategy using optimal dosage of beta blockers with/without ivabradine on the onset of acute adverse events in DCM-DMD patients.
Methods: We retrospectively analysed all patients aged >9 years with DCM-DMD and systolic dysfunction (ejection fraction <45%) referred to our institution, treated with a HRR strategy. Our institutional protocol established to uptitrate beta blockers with ivabradine in order to reach the target HRR of 20% or reaching 60 beats/min every two weeks [group 1]. These data were compared to control group, not receiving the HRR strategy [group 2]. We evaluate if the HRR strategy had an effect on: i. left ventricular ejection fraction, measured by echocardiography at baseline, and during follow up; ii. reduction on the rate of death and/or new hospitalizations ii.
Results: From our database of 80 DCM-DMD patients, 20 patients matched all entry criteria (mean age 19.6 + /- 5.7 yrs; group 1: $n = 13$; group 2: $n = 7$) and clinical, echocardiographic data were collected during follow up 2.2 ± 1.1 yrs.. In the group 1, mean HR was 95 ± 12 bpm at baseline, during follow up decreased to 62 ± 7 bpm ($p < .001$) while in the group 2 mean HR was 87 ± 13 bpm at baseline with no significant variation during follow up (mean 91 ± 10). HRR strategy showed left ventricular function improved (from baseline EF $28 \pm 3.2\%$ to 35 ± 4.2 EF%). Adverse events occurred in 15,4% vs 85,7% in the group 1 and group 2, respectively ($p < 0.0001$).
Conclusion: HRR strategy seems to be effective in reducing the incidence of acute adverse events and improving ventricular function in DCM-DMD patients.

O2873 - OUTCOME OF PAEDIATRIC PATIENTS WITH CONGENITAL HEART DISEASE TRANSFERRED FROM THEIR REGIONAL CARDIAC INTENSIVE CARE UNIT TO A UNIT WITH TRANSPLANT AND MECHANICAL SUPPORT CAPABILITY

Barbara Cardoso[1], Emma Simpson[1], Lee Ferguson[1], Judit Llevadias[1], Teresa Chilà, Margarita Guillèn[1], Yamuna Thiru[1], Fabrizio De Rita[2], Asif Hasan[2], David Steven Crossland[3]
Freeman Hospital, Department of Paediatric Intensive Care, Newcastle Upon Tyne-United Kingdom[1]; Freeman Hospital, Department of Cardiothoracic Surgery, Newcastle Upon Tyne-United Kingdom[2]; Freeman Hospital, Epartment of Paediatric Cardiology, Newcastle Upon Tyne-United Kingdom[3]

Background: Children with congenital heart disease (CHD) admitted to a Paediatric Cardiac Intensive Care Unit (PCICU) who exhaust conventional therapy or surgical options may be transferred to a PCICU with transplant and mechanical support capability for further management. We aimed to describe the management and outcome of this group- not all of whom are referred 'for transplant'.
Materials and Methods: Retrospective analysis of the records of patients with CHD and circulatory failure transferred from their regional PCICU between January 2011 and January 2016. Patients with cardiomyopathy were excluded.
Results: Twenty-seven patients were transferred and overall survival was 78% at 1 month and 61% at 1 year. 59.3% were male, median age 9.8 months IQR 4.4–44.6 months and 11 (41%) had univentricular physiology. Eight (29.6%) were transferred on ECMO. Seventeen patients were listed for cardiac transplant: 12 transplanted, 5 deaths on waiting list, 2 post-transplant deaths (day 102 and 109). Six VADs were undertaken as bridge to transplant (3 transplanted, 3 deaths onlist). Six patients were managed conventionally (4 further surgery, 2 medical/ ECMO only), all survived. Four patients were considered unsuitable for any further intervention including transplant. There was no difference in mortality between univentricular and biventricular circulations

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(p = 0.346) or those transferred on ECMO vs non-ECMO (p = 0.476). Survival was lower at <1 year of age compared to >1 year (survival at 30 days 64% v. 92%; one year 44% v. 77%, p = 0.051).
Conclusions: Although transplant is effective for CHD patients transferred acutely to a quaternary PCICU, mortality on the waiting list is a serious issue, partially due to limited mechanical support options for these patients. A carefully selected sub-group can be managed successfully with medical therapy and conventional surgery and this should be an integral part of the management offered by such PCICUs.

O2961 - LACK OF REJECTION AND EVIDENCE OF AUTOLOGOUS CELL REPOPULATION FOLLOWING HETEROTOPIC IMPLANTATION OF DECELLULARIZED PULMONARY HOMOGRAFTS. PATHOLOGICAL RESULTS IN A MURINE MODEL

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Objective: Tissue guided regeneration by means of decellularized scaffolds is based on the repopulation principle by the recipient autologous cells. Aim of this study was to evaluate inflammatory and regenerative responses after heterotopic implantation of decellularized pulmonary valve homografts (PVH) in a murine model.
Methods: Six PVH, harvested from wild-type Sprague-Dawley (SD) rats, underwent decellularization protocol consisting of 3 cycles of detergent-enzymatic treatment (DET) constituted by sodium deoxycholate (SDC) and DNase-I. Six native, undecellularized PVH served as control. Four decellularized and 4 native PVH were implanted in the abdominal aorta of green fluorescent protein (GFP) SD rats; the 4 unimplanted PVH, both native and decellularized served as decellularization control. Grafts, explanted 15 and 30 days after surgery, underwent gross, X-ray, histology, immunohistochemistry, electron microscopy immunogold labelling with the antibodies: anti-Von Willebrand factor (vWF), Smooth Muscle cell Actin (SMA), Vimentin (VIM), CD45, CD68 and anti-GFP. Cell density/mm2 was also measured.
Results: Before decellularization, PVH showed repopulation endothelial lining, valve interstitial cells as well and muscle cells in lamellar units. After decellularization, PVH exhibited complete decellularization (A), with DNA content of 5 ng/μl (removal greater than 95%). After 15 and 30 days from implant, native PVH showed severe cellular rejection, while decellularized PVH exhibited SMA positive myointimal cell layer, cell repopulation in outer media wall (B, C) and partial endothelial lining, in the absence of rejection. Cell density/mm2 was 126.14 ± 69.29 vs 132.07 ± 30.00 (p = NS) in the decellularized PHV media 15 and 30 days after implant, respectively while in the native was 533.72 ± 95.48 (p < 0.01). In the decellularized PVH wall, CD45 positive large, undifferentiated cells were found. All repopulation cells were GFP positive (D).
Conclusions: In this murine model, an effective removal of valve and arterial wall cells was obtained by DET-SDC decellularization treatment. Autologous GFP positive cell repopulation occurred with autologous cell origin evidence and no rejection.

O3026 - CARDIAC REGENERATIVE THERAPY USING CARDIAC PROGENITOR CELL FOR HYPOPLASTIC LEFT HEART SYNDROME AND SINGLE VENTRICLE.

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Backgrounds: Although the surgical result of hypoplastic left heart syndrome(HLHS) and single ventricle(SV) have improved in recent era, patients with SV, especially right dominant SV are particularly at risk for heart failure accompanied by high mortality We conducted cardiac regenerative strategy using cardiac progenitor cell infusion in children with HLHS and SV since 2011. The aim of this study is to determine whether intracoronary delivery of autologous progenitor cell improves cardiac function in patients with HLHS and SV.
Methods: Between January, 2011, and March, 2015, 41 children (2.8 ± 1.4 yr) with HLHS and SV were assigned to receive intracoronary infusion of cardiac progenitor cells after staged palliation. Of these, 17 patients were initially assigned to the control group. All received late CDC-infusion as an alternative option at a 3-month follow-up examination, and completed 3-month to 1-year follow-up studies after protocol intervention. Cardiac tissues were harvested during bidirectional Glenn procedure in 10 and Fontan in 31 patients. Progenitor cells were isolated and cultured. Progenitor cell was selectively infused into each coronary artery 1-2 months after surgery.
Results: There were no major adverse events up to 2 years. Based on cMRI, children received progenitor cell infusion showed a significant improvement of ventricular ejection fraction (Δ Ejection fraction- (%) :1.3 vs 6.4), ventricular mechanical efficiency (Ea/Ees; systemic ventricle and ventriculoarterial coupling) (ΔEa/Ees : 1.6 vs 1.2) and ventricular stiffness(Δ Ventricular stiffness:0.18 vs 0.12). In addition, plasma BNP levels and heart failure status were all markedly improved during follow-up observation
Conclusion: Intracoronary infusion of cardiac progenitor cells has improved ventricular function, ventricular mechanical efficiency, and ejection fraction and ventricular stiffness.

O3027 - THE PUMPKIN TRIAL STUDY DESIGN AND RATIONALE

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Background: Severe heart failure in children carries high mortality. Durable ventricular assist device (VAD) options for smaller children are currently limited to pulsatile devices that carry a high risk of stroke and lack portability. The Jarvik 2015, a fully implantable axial flow pump, similar in design to contemporary adult

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devices, has emerged as a promising treatment alternative for bridge-to-transplant support in smaller children.
Methods: The PumpKIN trial is an Investigational Device Exemption (IDE) study designed to evaluate the Jarvik 2015 VAD in children to support regulatory review of the device under the HDE regulation. The IDE study, which was conditionally approved by the FDA in 2016, is set to begin enrollment in 2017.
Results: The PumpKIN trial is a randomized clinical trial comparing the safety and probable benefit of the Jarvik 2015 VAD to the Berlin Heart EXCOR® Pediatric VAD. Children between 8 and 20 kilograms with 2-ventricle circulations and class IV heart failure (INTERMACS profile 1 or 2) despite optimal medical therapy and candidates for heart transplant will be enrolled. The primary probable benefit endpoint is the survival to transplant or 180 days of support without severe neurological dysfunction or randomized device failure. The primary safety endpoint is the incidence of protocol-defined adverse events using PEDIMACS criteria (version 5.0). The study is powered to show non-inferiority of the primary safety endpoint and will enroll a total of 88 subjects randomly assigned in a one-to-one ratio to each study arm.
Conclusions: The PumpKIN trial will be the first randomized clinical trial of a pediatric ventricular assist device. It is anticipated that data generated from the study will provide critical safety and effectiveness information for the first axial flow pump designed specifically for children with advanced heart failure.

O3031 - A NOVEL MINIATURE IMPLANTABLE CONTINUOUS FLOW PEDIATRIC VENTRICULAR ASSIST DEVICE

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Continuous flow ventricular assist devices (CFVADs) are demonstrated significant improvements in survival and quality of life in adults with heart failure. Unfortunately, no CFVAD designed for use in children and infants is available. We are developing a miniature blood pump specific to the pediatric heart failure population, based a miniature platform blood pump technology.
Methods: The pump is the size of a 'AA' battery with a non-contacting bearing and was designed for placement directly into the left ventricle with blood flow directed through an 8-10 mm graft anastomosed to the aorta. The pump generates 0.5–4 LPM of blood flow against pediatric systemic pressures. Hydraulic and hemolysis performance were evaluated in vitro. Anatomic fit was assessed using imaging from children from 2.5–25 kg imported into Mimics (Materialise, Plymouth, MI). Acute and chronic implants were performed in sheep for durations up to 3 months. Hemocompatibility was assessed by blood chemistry, plasma free hemoglobin (PFH), flow cytometric assays of platelet and leukocyte activation, aggregation, and microparticles, and by von Willebrand Factor function and distribution.
Results: From anatomic studies, intraventricular pump placement is acceptable for children <10 kg, potentially down to 3 kg, depending on ventricular dilation. In vitro hemolysis testing demonstrated low hemolysis (NIH <0.08). During acute and chronic implant testing in 26–47 kg sheep, 0.8–4.2 LPM was produced with low hemolysis (<20 mg/dL). There was no sustained increase in platelet or leukocyte activation or aggregation post-implant, and VWF function was preserved, in spite of no post-operative anticoagulation. Renal infarcts were only observed in 1 of 10 chronic implants in an animal with profound hypertension.
Conclusions: These promising data confirm the feasibility of using a miniature CFVAD to support children and infants with heart failure while achieving improved hemocompatibility.

POSTER ABSTRACTS

ADMINISTRATIVE

P1107 - SAFETY NUMBERS IN THE CARDIAC INTENSIVE CARE UNIT (CICU)

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Background: The availability of an efficient electronic event reporting system intended to increase the number of reported events in CICU. This system coupled with other programs strengthens and facilitates reporting.
Objective: The CICU Safety Council and unit leadership aim to increase the CICU's safety event reporting through identification of barriers and development of programs to encourage reporting.
Methods: The council and leadership advocated to improve the electronic safety event reporting system by implementing various strategies to overcome identified barriers. These include: improvement of the RL system, provision of access to council members, multiple campaign methods such as email, nurse huddle announcements, person to person feedback, and the selection of a monthly winner for a good "catch". Also implemented are: transparency and the utilization of the Just Culture Algorithm, peer accountability and peer coaching, mini-apparent cause analysis, monthly reports through blame-free emails and monthly staff meetings.
Results: A significant increase in reported safety events in the CICU were observed when comparing the two most recent years. There were 473 events reported during the FY 2016 compared to 347 events reported in 2015.
Conclusion: Increase in the number of safety event reporting was achieved through strong collaboration between CICU leadership and the Safety Council and of various strategies and programs applied.

P1211 - KNOWLEDGE INTERVENTION AT CAMP FOR CHILDREN WITH CHD

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Transitioning from pediatric care to adult care can be difficult for patients as they must obtain responsibility for their healthcare. Many adolescents and adults with congenital heart defects (CHD) are unable to name their diagnosis and have inadequate knowledge of treatment and prevention of cardiac complications, such as knowledge about endocarditis, effects of smoking and alcohol and contraception options. Successful transition is associated with improved medical and psychosocial outcomes in adults with

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congenital heart disease, as well as decreasing cost of medical care. Specialized camps targeted at children with chronic conditions have shown to be psychosocially beneficial. However, no research focuses on the benefits of a knowledge intervention of children with CHD in a camp setting. Many adolescents with congenital heart defects (CHD) have inadequate knowledge of treatment and prevention of cardiac complications, but will need such knowledge for successful transition to adult care. We will test knowledge improvement after a camp-based educational intervention. We were particularly interested in level of knowledge about endocarditis among our campers. We also aimed to assess camper satisfaction and feedback.35 youth with CHD age 10-14 attended a 5-day sleepover camp with medical staff and peers. Camp included group and individual education targeting knowledge about diagnosis, management/prevention of complications, and endocarditis. Youth completed a cardiac knowledge measure (modified version of the Leuven Knowledge Questionnaire) before and after camp. Youth demonstrated a significant improvement in knowledge about CHD diagnosis and prevention of complications, with particularly large gains in knowledge about endocarditis. There was no change in knowledge about sexuality and contraception, and these areas had a relatively low percentage of correct responses. CHD education is feasible and acceptable to parents and youth in the camp setting. Education in this setting may facilitate transition goals, but knowledge about sexuality may require tailored intervention for youth with CHD.

P1290 - GOAL ALIGNMENT IS CRITICAL TO THE SUCCESS IN CONGENITAL CARDIAC SURGERY TEAMWORK AND COMMUNICATION ARE THE CORNERSTONE

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Background: To evaluate whether technical error, inadequate teamwork and communication in congenital cardiac surgeries result in flow disruptions, which can pose serious risk to patients, and to improve teamwork to improve patient safety.
Methods: Qualitative interview study was conducted with the congenital heart surgery team. Members of the surgical team were interviewed about the following four themes: general processes, goals, communication and leadership. All responses were grouped into the 20 categories, and were analyzed further for the frequency of relevant quotes in each category, by the interviewees.
Results: All the participants agreed that the surgeon oversees the OR, 60% acknowledge that leadership can change at different points in the operation, >70% mentioned the following four categories: (1) patient-centered goals, (2) primary vs. secondary goals, (3) leadership and (4) conflict resolution. 80% of the participants mentioned the safety of the patient as their primary goal, while 70% of the staff mentioned having primary and secondary goals, and describing that secondary goals conflict amongst individuals. 60% believed having a diverse team creates better care by providing a broader range of treatment options and expertise, 50% mentioned "new team" leading to tensions amongst members. 70% of participants mentioned communication as the primary method of conflict resolution. This unit employed different communication methods: pre-huddle, post-huddle, read- backs and checklists. Condescension was mentioned by 50%, which threatens the efficiency of the team. Frequency of responses that mentioned goals, conflict, teamwork, communication, and learning were 23%, 24%, 23%, 13%, and 17%, respectively.
Conclusions: While conflict is an inherent part of the OR because everyone's secondary goals are profession specific, conflict must be addressed and reduced through improved communication to maximize efficiency and improve patient safety. Developing methods of constructive communication that allows each team member to define their primary and secondary goals is vital.

P1405 - GASTROINTESTINAL BLEEDING IN PATIENTS AFTER FONTAN OPERATION

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Background: Gastrointestinal (GI) bleeding is a rare but potentially life-threatening complication in patients after Fontan operation. However, the clinical presentation and risk factors of GI bleeding remain poorly understood.
Methods: We retrospectively reviewed the medical records of 117 patients who received follow-up in our institute between January 2000 and March 2016. Major GI bleeding is defined as fatal, hemodynamic instability, symptomatic, decrease in hemoglobin ≥ 2 g/dL over 24 hour or requiring transfusion.
Results: Nine patients (7.7%) had major GI bleeding and all of them underwent endoscopic examination. Eight of 9 patients had received thromboprophylaxis therapy, including warfarin in 7 patients. Median interval between the onset of GI bleeding and Fontan completion was 2.3 years, ranging from 10 days to 12 years. We categorized these patients into 2 groups: early-onset (≤3 months, n = 2) and late-onset (>3 months, n = 7). Both patients with early-onset GI bleeding are associated with sepsis and multiple organ dysfunction. Of 7 patients in late-onset group, 5 (71%) patients had clinical presentation of protein-losing enteropathy (PLE); 6 (86%) patients had INR > 2 (all under warfarin) during first GI bleeding episodes; and 7 (100%) patients had significant obstruction of Fontan pathway or stenosis of pulmonary arteries. The mortality rate is much higher in early-onset group (100%, 2/2 vs 29%, 2/7). Among them, one in late onset group developed significant GI bleeding complicated with Fontan failure and subsequently death due to brain edema and herniation. Massive GI bleeding with concurrent intracranial hemorrhage lead to the demise of the 3 remaining patients.
Conclusion: After Fontan completion, patients with early-onset GI bleeding are associated with high mortality rate. The causes of late-onset GI bleeding maybe multifactorial, including bleeding tendency and increased systemic venous pressure due to obstruction of Fontan pathway. Late-onset GI bleeding may be a part of the clinical spectrums of PLE.

P1512 - PRACTICE MAKES PERFECT IMPROVING TEAM COMMUNICATION WITH TEAM BASED SIMULATION

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Background: Development of a culture of open communication centered on goal alignment is critical to ensure safe and effective care. Team training that simulates an entire surgical scenario from preoperative preparation to surgical intervention to recovery ensures the team can respond effectively and efficiently. Team based training is used extensively in other industries like aviation and nuclear power.
Methods: From October 2013 to September 2015, we have completed four in-depth congenital heart team scenarios that are geared to the entire operative day from patient entering the operating room to operative procedure to transfer to the intensive care unit to team handoff to intensive care scenarios. allowing for goal alignment and team-based decision making over the entire continuum of the patient care team. After completing the simulation, the entire team meets to analyze the day and to identify communication failures, workflow disturbances, and equipment issues.
Results: Over the 2 years, the use of the in-depth, intensive simulations we have developed several dynamic communication tools and methods to avoid conflicts, following the American Heart Association's guidelines. We have seen a reduction in blood draws, complications like infections.
Conclusions: In the operating room and the ICU, we can develop team-based learning and develop a culture of communication centered on goal alignment. We can change scenarios and introduce very real, if infrequent, disasters. This unique environment tests the team's response as a cohesive unit. We share the same primary goal of the safe return of the patient to the family; however, each group has sub-goals and varying opinions on how to attain the primary goal. During the simulation, we can test different aspects of communication; teams with excellent goal alignment and understanding will respond to significant events in a timely and smooth manner, decreasing possible negative effects to the patient.

P1552 - THE BIRTH PREVALENCE AND RISK FACTOR OF CONGENITAL HEART DISEASE (CHD) IN INFANTS BORN FOLLOWING ASSISTED REPRODUCTIVE TECHNOLOGY(ART)

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Background: The use of assisted reproductive technology (ART) to treat infertility is increasing rapidly. Although the growing number of infants born following ART, there is lack of data about the prevalence of congenital heart defect and the risk factors of CHD in these neonates. We conducted this study to assess the association between CHD and ART.
Method: The medical records of mothers who were pregnant with ART in Asan Medical Center, since 2005 and their infants were reviewed retrospectively. The echocardiographic study was done in neonatal period for the evaluation of cardiac murmur and abnormal result of fetal echocardiography. CHD was defined as structural abnormality of the heart or intrathoracic great vessels with actual or possible functional significance. We assumed maternal risk factors for CHD as age, gestational age at birth, multifetal gestation, structural anomaly of uterus, parity, thyroid dysfunction, diabetes mellitus and hypertension.
Result: Three hundreds and sixty mothers and 484 infants were enrolled and 36 infants (7.4%) were born with CHD; 16 treatment required patent ductus arteriosus (PDA) (3.3%), 17 atrial septal defects (ASD) (3.5%), 9 ventricular septal defects (1.9%), 1 pulmonary valve stenosis, 1 aortic valve stenosis, 1 transposition of great arteries and 1 Ebstein's malformation. After excluding PDA and ASD, 12 patients (2.3%) had complex-CHD (cCHD). Among the possible risk factors for cCHD, structural anomaly of uterus (Odds Ratio [OR], 2.950, p = 0.070 ; 95% CI 0.916-9.506) and hypothyroidism (OR, 2.151, p = 0.343 ; 95% CI 0.441-10.479) increase the prevalence of cCHD.
Conclusion: The birth prevalence of CHD in neonates born with ART was increased in this study. However, there's a limitation as we could not figure out the proportion of fetal termination and the exact cause of termination. The multi-centered large scale study for the association between ART and CHD should be performed.

P1616 - THE FETUSES WITH EBSTEIN ANOMALY OR TRICUSPID VALVE DYSPLASIA A SINGLE CENTRE EXPERIENCE

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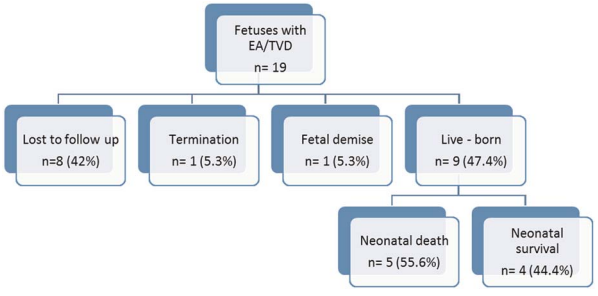
Ebstein anomaly and tricuspid valve dysplasia are rare congenital tricuspid valve malformations associated with high perinatal mortality. In this report, we review 19 fetuses diagnosed Ebstein anomaly or tricuspid valve dysplasia from 2013 to 2016.
Methods and Results: Twelve of the patients were tricuspid valve dysplasia (TVD) and seven of them were Ebstein anomaly (EA). The mean gestational age at the diagnosis was 29,7 + 6,3 (18–38) weeks. All of them had right atrial dilatation and cardiomegaly. Cardiothoracic area (CTA) ratio was 0,46 + 0,17 (0,31–0,73). Thirteen fetuses had severe tricuspid regurgitation (TR), six of these fetuses had retrograde duct flow and three fetuses had pulmonary regurgitation (PR). Two fetuses had hydrops fetalis. Three fetuses had small ventricular septal defects, one fetus had pulmonary atresia and one noncompaction. Among all patients, there were 8 lost to follow-up, 1 fetal demise and 1 termination. Nine fetuses were live-born but five of them died first days of the life. The mean CTA ratio was higher in neonatal death group than in the living patients (0,54 (0,44–0,73), 0,44 (0,31–0,61) respectively). In neonatal death group; two patients had comorbid diseases (diaphragm hernia and bilateral renal agenesis), one patient who had no antegrade pulmonary valve flow underwent to shunt operation and died with sepsis. Other two patients died within 24 hours of life with reduced cardiac output and acidosis. Four patient are following up with mild – moderate TR. These patients ages are between 3 months and 2,5 years.
In conclusion; despite major advances in prenatal care and the diagnosis and management of congenital heart disease over the past several decades, perinatal mortality in fetuses with EA/TVD remains alarmingly high.



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P1624 - APPLYING A CLINICAL SERVICE RE DESIGN METHODOLOGY TO ESTABLISH A STATEWIDE MODEL OF CARE TO SUPPORT THE DEVELOPMENTAL NEEDS OF CHILDREN WITH CONGENITAL HEART DISEASE FOLLOWING EARLY OPEN HEART SURGERY

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Background: Each year 100 Queensland infants with congenital heart disease (CHD) require open-heart surgery before 12 months of age, increasing their risk of poor neurodevelopmental outcomes. Developmental surveillance is recommended for infants following neonatal open-heart surgery to mitigate risk through early intervention. The Queensland Paediatric Cardiac Service (QPCS) Neurodevelopmental Follow-up Program offers robust developmental screening limited to specific high-risk cohorts and centralised to the quaternary hospital, posing challenges within a large geographical state. This project aims to establish a holistic, statewide approach to optimising developmental outcomes of Queensland children with CHD.
Methods: A collaborative partnership was established between QPCS and the Queensland Child and Youth Clinical Network to maximise statewide engagement and project outcomes. Funding was provided by the Healthcare Improvement Unit. Partnership with Innovation Change and Redesign Excellence program (iCARE) enabled application of a robust, clinical service re-design methodology, including collaboration with three hospital and health services (HHSs), and executive endorsement. Stakeholder consultation, including consumer engagement, throughout the planning and diagnostic phases established the platform from which solution and implementation phases could launch.
Results: Over 60% of infants who undergo open-heart surgery in the first 12 months of life live outside the Brisbane metropolitan area, with few linked into local supports and services. Consultation with consumers and services providing developmental supports identified barriers to optimising the developmental outcomes of this cohort. Key themes identified during the diagnostic phase include; improving communication processes, empowering parents, information and resource sharing, capability building within services, and the need for centralised data collection.
Conclusions: Application of this methodology has enabled rigorous planning and effective stakeholder engagement across partner sites, ensuring contextualised project outcomes that meet the needs of families within each HHS. It is anticipated that this methodology will support application of project outcomes across other Queensland HHSs, and to the broader CHD population.

P1643 - SERUM CREATININE LEVELS IN THE FIRST 7 DAYS OF LIFE IN NEONATES WITH CONGENITAL HEART DISEASE

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Background: Acute kidney injury after cardiac surgery is an important complication. The presence of maternal creatinine and daily changes in neonatal renal function make it indefinable in neonates. We aimed to examine changes in serum creatinine level over the first week of life in neonates with congenital heart diseases (CHDs).
Materials and methods: The clinical data of 159 consecutive neonates with CHD and 125 neonates as controls were retrospectively reviewed. The exclusion criteria were birth weight of <2 kg and other major complications such as intracranial and abdominal lesions, congenital kidney diseases, and congenital diaphragmatic hernia. The neonates were classified into 3 groups, namely the simple and complex CHD with bi-ventricle groups (sBV and cBV, respectively) and single-ventricle group (SV). Follow-up was completed when an invasive procedure was performed. Serum creatinine levels over the first 7 days were compared.
Results: No significant differences in birth weight and gestational age were found between the CHD and control groups. The mean serum creatinine levels in the two groups were similar over the first 48 hours. Creatinine level kept increasing during 2 days after birth in both groups (CHD vs control group: 0.79 ± 0.14 vs 0.83 ± 0.16 mg/dl at 2 days of age, $p > 0.05$), and decreased in the control group. However, the CHD group maintained a higher serum creatinine level for 7 days after birth (0.59 ± 0.17 vs 0.36 ± 0.08 mg/dl, $p < 0.01$). No significant differences were found in the chronological changes in serum creatinine level among the 3 groups of CHD neonates (sBV, cBV, and SV).
Conclusions: Serum creatinine level was significantly higher in the CHD neonates than in the healthy neonates during the first week of life. Cardiac morphological differences seem to have little influence on the serum creatinine levels of neonates.

P1804 - EVALUATION OF CHILDREN HAVING CONGENITAL HEART DISEASE AND HOSPITALIZED WITH THE DIAGNOSIS OF LOWER RESPIRATORY TRACT INFECTION

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Lower respiratory tract infections (LRTI) have an important mortality and morbidity for children with congenital heart disease (CHD). In this study, 50 children who have congenital heart disease and hospitalized with the diagnosis of LRTI are analyzed in Children's Hospital of Ege University Medical Faculty. 50 pediatric patients who have CHD and hospitalized with LRTI in cardiology service between 2013 and 2015 were studied. For these patients who were previously diagnosed or have recently been diagnosed with CHD, clinical symptoms, acute phase reactants, chest X-ray and nasopharyngeal swab in respiratory virus panel (Multiplex Polimerase Chain Reaction) evaluated. Patients were classified into two groups as cyanotic and acyanotic. The groups were examined in terms of age, gender, RSV prophylaxis, hospitalization time, causative pathogen.Results: Of the 50 cases, 16(% 32) were diagnosed with cyanotic CHD and 34 (%68) were diagnosed with acyanotic CHD. Boy/girl was in ratio of 26/24. 9 cases (%18) were diagnosed with VSD and ASD, 7 cases (%14) were diagnosed with only VSD, 6 (%12) cases were diagnosed with DORV and 1 case were diagnosed with PDA + ASD. All other cases had mixed type diagnostics including VSD. The average age of patients diagnosed with cyanotic and acyanotic CHD, were $23,88 \pm 28,81$ months, $12,25 \pm 15,45$ months old respectively. %50 of patients diagnosed with cyanotic CHD and % 73.5 of patients diagnosed with acyanotic CHD were less than 12 months old. Hospitalizations most frequently occured in winter (17 cases, %34), then secondly in spring (14 cases, %28). In 3 of patients diagnosed with cyanotic CHD (%18.75) and 6 of patients

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diagnosed with acyanotic CHD (%17.64), viral agents were detected. LTRI and specifically RSV pneumonia are important causes of mortality and morbidity in patients diagnosed with CHD

P1886 - EFFECTIVENESS OF CARDIOVASCULAR SCREENING IN 1 065 097 SCHOOL AGED CHILDREN

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Background: Undetected heart disease is one of the most common cause of sudden death in children. We conducted cardiac screening in school age children to detect the heart diseases.
Methods: From 1999 to 2014, a citywide cardiovascular survey of elementary school students had been performed in Taipei. A questionnaire regarding past history of heart disease, family history and cardiac symptoms was used. A 4-lead computerized electrocardiogram (ECG) & phonocardiogram were performed in each child. Patients with any abnormalities above were referred to hospital for final diagnosis.
Results: Totally 1,065,097 first grade of elementary school children underwent cardiac screening. Congenital heart disease or other types of heart diseases was identified in 5,416 children (5.1 /1000) of whom 401 children had not been diagnosed before. Atrial septal defect was the most common undiagnosed congenital heart disease, accounting for 265 children (66%). Complete heart block, WPW syndrome & prolonged QTc interval were found in 14, 818, and 99 children, respectively. Cardiomopathy was diagnosed in 72 children of whom 19 children had not been diagnosed.
Conclusions: Cardiac screening in school-aged children using questionnaire, ECG & phonocardiography is effective. Atrial septal defect was the most common undiagnosed congenital heart disease. Many kinds of cardiac conduction disorders and even life-threatening condition can also be identified through this cardiovascular screening.

P1988 - MULTI CENTER CONGENITAL HEART DISEASE DATABASES ACTIVE WORLDWIDE

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There are at least 39 known multi-center efforts collecting information on patients with congenital heart disease that we have identified worldwide. Some of these are geographically limited while others are very disease or specialty specific. We have collated and present here all of the currently active databases and registries. These were identified by personal involvement, word of mouth, PubMed searches and Google(TM) searches utilizing terms such as "cardiac, congenital, surgery, intensive care, critical care, cardiology, databases, and registries" in various combinations. Hospitals in developed countries may spend millions of dollars in participation fees and indirect expenses (salaries, travel). There needs to be better coordination between existing registries and databases and mechanisms for cross-linking data between them. Access to these databases also needs to be made more usable for resource-poor countries and for "mission" trips. Prior to starting more databases or registries it would be useful for planners to see if existing ones might suit their purposes.

Table.

ACTIVE Clinical Databases and Registries	Acronym	Sponsoring Organization and/or Data Center	Target Patient Population
Australia and New Zealand Fontan Registry		Australia and New Zealand Congenital Heart Research Centre	All patients in Australia and New Zealand who have undergond a Fontan-type single ventricle repair
Canadian Outcomes Registry Late After Tetralogy of Fallot Repair	CORRELATE	Canadian Institutes of Health Research/The HUB-University of Toronto	All patients in Canada who have undergone surgical repair of Tetralogy of Fallot, age > 12 yrs
Congenital Evaluation, Reporting, and Tracking Endeavour	CONGENERATE	Adult Congenital Heart Association/McGill University, University of Sherbrooke, University of Montreal	Adult patients with Congenital Heart Disease/ Defects
Congenital Cardiac Anesthesia Society Database	CCAS	Congenital Cardiac Anesthesia Society/Duke Clinical Research Institute	Any procedures involving congenital cardiac patients, limited to STSCHSD participating sites
Congenital Cardiac Catherizations Project on Outcomes	C3PO	Children's Hospital Boston	Patients undergoing congenital cardiology interventional procedures
Congenital Cardiovascular Interventional Study Consortium	CCISC	Children's Hospital of Michigan	Interventional cardiology procedures, tends to be very lesion specific
Congenital Heart Surgeons Society Database	CHSS	Congenital Heart Surgeons Society Data Center at The Hospital for Sick Children, Toronto	Specific Congenital Heart Lesions depending upon study area (currently 4 active studies (LVOTO, AVSD, AAOCA, Tricuspid Atresia; one pending, Ebsteins)
European Congenital Heart Surgeons Association Database**	ECHSA	European Congenital Heart Surgeons Association	Patients undergoing any ongenital heart surgical procedure
European **R**egistry for **I**CD and CRT Devices in Pediatrics and Adults with Congenital Heart Disease	EURIPEDES	Association for European Paediatric and Congenital Cardiology/European Society of Cardiology/Competence Network Congenital Heart Defects	Pediatric and Congenital Patients with ICD or CRT devices
European Registry for Patients with Mechanical Ciruculatory Support	EUROMACS	European Association of Cardio-Thoracic Surgery	All age patients with durable and temporary mechanical support devices
Extracorporeal Life Support Organization Registry	ELSO	Extracorperal Life Support Organization	Patients undergoing extracorporeal membrane oxygenation (ECMO)
German Heart Institute Myocarditis Registry for Children	MYKKE	Association for European Pediatric Cardiology/Competence Network for Congenital Heart Defects	Pediatric myocarditis
Guangdong Registry of Congenital Heart Disease			All patients with a diagnosis of CHD
Improving Pediatric and Adult Congenital Treatments	IMPACT	American College of Cardiology/ National Cardiovascular Data Registry/Mid America Heart Institute	Patients undergoing congenital cardiology interventional procedures

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Table. *Continued*

ACTIVE Clinical Databases and Registries	Acronym	Sponsoring Organization and/or Data Center	Target Patient Population
Interagency Registry for Mechanically Assisted Circulatory Support	INTERMACS/ Pedi MACS	International Society for Heart and Lung Transplantation/ University of Alabama	Patients with durable and temporary mechanical support devices
International Fetal Cardiac Intervention Registry	IFCIR	Multiple sponsoring organizations	All patients undergoing fetal cardiac interventions
International Pediatric Heart Failure Registry	iPHFR	International Society for Heart and Lung Transplantation/ University of Alabama	Congenital and Acquired Heart Failure in Children
International Quality Improvement Collaborative for Congenital Heart Surgery in Developing World Countries	IQIC	Global Forum in Humanitarian Medicine for Cardiology and Cardiac Surgery	Low resource/ developing countries undergoing CHS
International Society for Heart and Lung Transplantation	ISHLT	International Society for Heart and Lung Transplantation/ University of Alabama	Pediatric Heart and Lung transplants
Japan Congenital Cardiovascular Surgical Database	JCCVSD	Japanese Society for Cardiovascular Surgery	All patients in Japan undergoing congenital cardiac surgery
Japanese Registry for Patients with Mechanical Circulatory Support	J-MACS		All age patients with durable and temporary mechanical support devices
National Anesthesia Clinical Outcomes Registry	NACOR	Anesthesia Quality Institute/ American Society of Anesthesiologists	All patients undergoing anesthesia in participating hospitals
National Congenital Heart Disease Audit		National Institute for Cardiovascular Outcomes Research (NICOR)	All patients in UK undergoing congenital cardiac surgery
National Pediatric Cardiology Quality Improvement Collaborative	NPC-QIC	Joint Council on Congenital Heart Disease/Cincinnati Children's Hospital Medical Center	single ventricle patients
National Register for Congenital Heart Defects		Competence Network for CHD	Pediatric and Congenital Cardiac Disease Patients
Paediatric Intensive Care Audit Network	PICANet		PICU admissions, including Cardiac patients
Pedatric Heart Transplant Study	PHTS	Pediatric Heart Transplant Foundation/University of Alabama	Pediatric Heart transplant recipients
Pediatric Cardiac Critical Care Consortium	PC4	Pediatric Cardiac Critical Care Consortium/University of Michigan	Cardiac ICU patients
Pediatric Cardiomyopathy Registry	PCMR	National Heart, Lung and Blood Institute (NHLBI)/Children's Cardiomyopathy Foundation	Patients with primary and idiopathic cardiomyopathy
Quebec Congenital Heart Database			Database derived from Administrative information on all patients in Quebec with a diagnosis of congenital heart disease
Research Registry of Pediatric Cardiac Surgery			All patients undergoing pediatric cardiac surgery in Finland
Scientific Registry of Transplant Recipients		Minneapolis Medical Research Foundation	Patients undergoing solid organ (Heart, Lung, Heart-Lung, Liver, Kidney, Pancreas, Intestine) transplants
STS Congenital Heart Surgery Database	STSCHSD	Society of Thoracic Surgeons/ Duke Clinical Research Institute	Patients undergoing any congenital heart surgical procedure
Swedish National Registry for Congenital Heart Disease	SWEDCON		All patients in Sweden with congenital heart disease
Swiss National Registry of Grown Up Congenital Heart Disease	GUCH	University Hospital, Basel, Switzerland	All patients with Adult Congenital Defects presenting in Switzerland
Tracking Outcomes and Practice in Pediatric PH	TOPP-2	Association for Pediatric Pulmonary Hypertension	Pediatric Patients with PH
UNOS	UNOS	United Network for Organ Sharing	Any patient receiving a solid organ transplant, including children
Virtual PICU Systems	VPS	Virtual PICU Systems/Children's Hospital Association	Pediatric ICU patients, including those with a cardiac diagnosis
Western Canadian Children's Heart Network Database		Western Canadian Children's Heart Network	All patients in Western Canadian provinces with CHD

ACTIVE Clinical Databases and Registries	Geographic Area	Website
Australia and New Zealand Fontan Registry	Australia, NZ	http://www.fontanregistry.com
Canadian Outcomes Registry Late After Tetralogy of Fallot Repair	Canada	http://www.hubresearch.ca/project-profiles/correlate/
Congenital Evaluation, Reporting, and Tracking Endeavour	US, Canada	http://www.congenerate.org
Congenital Cardiac Anesthesia Society Database	US, Canada	http://www.ccasociety.org/
Congenital Cardiac Catherizations Project on Outcomes	US	https://c3po-qi.chboston.org/
Congenital Cardiovascular Interventional Study Consortium	Worldwide	http://www.ccisc.net
Congenital Heart Surgeons Society Database	US, Canada	http://www.chssdc.org/
European Congenital Heart Surgeons Association Database**	Worldwide	http://echsacongenitaldb.org/
European **R**egistry for **I**CD and CRT Devices in Pediatrics and Adults with Congenital Heart Disease	Europe + Russia	http://www.euripides-registry.eu/
European Registry for Patients with Mechanical Ciruculatory Support	Europe + Russia	http://www.euromacs.org
Extracorporeal Life Support	Worldwide	http://www.elso.org

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Table. *Continued*

ACTIVE Clinical Databases and Registries	Geographic Area	Website
Organization Registry		
German Heart Institute Myocarditis Registry for Children	Germany	http://mykke.de
Guangdong Registry of Congenital Heart Disease	Guangdong Province, China	
Improving Pediatric and Adult Congenital Treatments	International	http://cvquality.acc.org/en/NCDR-Home/Registries/Hospital-Registries.aspx
Interagency Registry for Mechanically Assisted Circulatory Support	US	https://www.uab.edu/medicine/intermacs/
International Fetal Cardiac Intervention Registry	Worldwide	http://www.ifcir.org/
International Pediatric Heart Failure Registry	Worldwide	https://www.ishlt.org/registries/iPHFR.asp
International Quality Improvement Collaborative for Congenital Heart Surgery in Developing World Countries	Worldwide	
International Society for Heart and Lung Transplantation	Worldwide	http://www.ishlt.org/
Japan Congenital Cardiovascular Surgical Database	Japan	
Japanese Registry for Patients with Mechanical Circulatory Support	Japan	
National Anesthesia Clinical Outcomes Registry	US	http://www.aqihq.org
National Congenital Heart Disease Audit	UK	https://nicor4.nicor.org.uk/chd/an_paeds.nsf/vwcontent/home
National Pediatric Cardiology Quality Improvement Collaborative	US	http://jcchdqi.org
National Register for Congenital Heart Defects	Germany	http://www.kompetenznetz-ahf.de/en/research/register-biobank/
Paediatric Intensive Care Audit Network	UK and Ireland	http://www.picanet.org.uk/
Pedatric Heart Transplant Study	Worldwide	http://www.uab.edu/medicine/phts/
Pediatric Cardiac Critical Care Consortium	US, Canada	http://www.pc4quality.org
Pediatric Cardiomyopathy Registry	US	http://www.childrenscardiomyopathy.org/site/registry.php
Quebec Congenital Heart Database	Canada (Quebec)	
Research Registry of Pediatric Cardiac Surgery	Finland	
Scientific Registry of Transplant Recipients	US	http://www.srtr.org
STS Congenital Heart Surgery Database	US, Canada	http://www.sts.org
Swedish National Registry for Congenital Heart Disease	Sweden	http://www.ucr.uu.se/swedcon/
Swiss National Registry of Grown Up Congenital Heart Disease	Switzerland	
Tracking Outcomes and Practice in Pediatric PH	Worldwide	https://www.peph-association.org/
UNOS	US	http://www.unos.org
Virtual PICU Systems	Worldwide	http://www.myvps.org
Western Canadian Children's Heart Network Database	Canada	http://www.westernchildrensheartnetwork.ca/

P2212 - HIGH MEAN PLATELET VOLUME IN CONGENITAL HEART DISEASE WITH PULMONARY HYPERTENSION

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Backgrounds: Because thrombosis is one of the significant problems in congenital heart disease, platelet activity in this situation is an important issue to concern. Mean platelet volume (MPV) is measurable in routine blood cell counts and reflects to platelet activity.
Purpose: To investigate the relationship between platelet activity and hemodynamic parameters including quantity of pulmonary blood flow and pulmonary arterial pressure.
Methods: From 2012 to 2015, 279 cardiac catheterizations for congenital heart diseases without surgery were examined. Quantity ratio of pulmonary and systemic blood flow (Qp/Qs), mean pulmonary arterial pressure (mPAP), and brain natriuremic peptide (BNP) were compared to MPV. Each parameter was analyzed by regression analysis.
Results: Age at catheterization was 0 to 76 years old (median 5.3). Each parameter range, average and standard deviation were followed as; MPV 7.5 to 12.5 fL (mean 9.3, SD 0.8), SpO2 72 to 100% (average 96.6, SD 3.9), Qp/Qs 0.4 to 7.1 (average 1.7, SD 0.74), mPAP 11 to 60 mmHg (average 19.1, SD 6.2), BNP less than 5 to 3608.1 pg/ml (average 38.6, SD 226.0). In regression analysis to MPV, Qp/Qs had significant but weak positive relation (R 0.16, $p = 0.01$). SpO2, mPAP and BNP had no significant relation. However in 34 cases whose mPAP is higher than 25 mmHg, MPV had significant negative relation to mPAP ($R = 0.53$, coefficient = 0.043, $p = 0.001$) and not significant but positive relation to Qp/Qs ($R = 0.34$, coefficient = -0.29, $p = 0.05$).
Discussion: Although there is no relationship between MPV and hemodynamic parameters in all of native congenital heart disease, cases with pulmonary hypertension had higher platelet activity as higher pulmonary arterial pressure and lower pulmonary blood flow. In cases with high pulmonary vascular resistance platelet might be activated by endotherial injury due to pulmonary hypertension.

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P2288 - PAEDIATRIC CARDIOLOGY TRAINING IN THE UK

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Introduction: Paediatric cardiology has experienced enormous advances since its conception to current practice, incorporating highly developed medical, surgical and radiological techniques allowing significantly improved prognosis from hours or days to near normal life expectancies. We identify several important factors facing paediatric cardiology training in Britain; including the new junior doctor's contract, Shape of Training review and the future of the speciality in the post 'Brexit' U.K.
Method: We identified several key issues that have been subject to debate within paediatric cardiology and medicine in the UK. We surveyed the opinion of paediatric cardiology trainees throughout the U.K regarding the effect this may have on training. We also suggest several possible consequences on the service that these may have
Conclusion: Paediatric Cardiology is a unique speciality. However, as a speciality in the UK, it faces several potential challenges for the future. "The shape of training" proposal, with the ambition to improving functionality within the NHS, does not take into account small complex paediatric specialties'. One area of concern is the lack of clinical experience in training needed to function within this speciality. Potentially compounding this is the effects of the junior doctor contract on working hours, reducing exposure to the speciality. More subjective feelings of lower staff morale and difficult working conditions in such an emotive speciality were also an area of concern. Finally, concerns following the E.U referendum on the detriment this may have on research, information sharing and clinical experience were also highlighted. We have drawn attention to several areas of concern for paediatric trainees that, if approached early, may delineate the future generations of paediatric cardiologists.

P2487 - DETECTING HIGH RISK AREAS FOR CONGENITAL HEART DISEASE IN NORTHEAST BRAZIL

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Background: Congenital heart disease (CHD) is one of the major causes of neonatal morbidity and mortality in children. In developing countries, however, it is often under-reported leading to poor documentation of its true incidence. The objective of this study is to describe the epidemiological profile of CHD in the state of Paraiba from the implementation of a pediatric cardiology network; and also, to list the impact from its spatial distribution and associated risk factors.
Methods: Ecological study with quantitative and exploratory approach using data from neonates diagnosed with CHD from Jan-2012 to Dec-2016 in Paraíba, northeast Brazil. Data analysis was exploratory and spatial. Risk maps were generated to compare CHD cases among municipalities. Choropleth maps where created to spatially portray the Risk Ratio (RR). Clusters of CHD were identified via spatial scanning method and sociodemographic indices were compared among areas.
Results: Exactly 146,422 neonates were screened in the period. The Network team diagnosed 1,951 CHD as compared to 68 reported by the Brazilian official statistic site. For the RR, 52 municipalities (23.31%) presented RR >1.5, and in 4 (1.79%) it was greater than 4.0. Conversely, 119 municipalities (53.36%) presented lower RR for CC (RR < 1.0). The Scan spatial methods identified various clusters for CHD in the state. These clusters were associated with areas of high socioeconomic inequalities.
Conclusions: The present study identified a much higher incidence of CHD, in Paraíba, than what has been reported in the official sites. It also pointed to clusters of CHD within the State and to novel risk factors. These findings can be used to develop public strategies to promote health and prevent disease.

P2607 - USEFULNESS OF SOLUBLE UROKINASE PLASMINOGEN ACTIVATOR RECEPTOR (SUPAR) AS AN INFLAMMATORY BIOMARKER IN OBESE CHILDREN

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Objective: Soluble urokinase plasminogen activator receptor (suPAR) has emerged as a relatively new biomarker that reflects increased inflammatory status and been associated with cardiovascular risk. We wanted to investigate the predictive value and usefulness of suPAR as an inflammatory biomarker in obese children.
Methods and Results: Of the total 136 participants, 76 (36 male, 40 female) were in obese group and 60 (24 male, 36 female) were in control group. The median age was 12.05 (6.16-17.30) years old for obese group, and 12.83 (8.00-16.75) years old for control group. Obese children had statistically significantly higher heart rate, systolic and diastolic blood pressure, EAT and LV mass than control group ($p < 0.01$). The median suPAR level in obese group was not statistically different than in control group (0.54 vs. 0.59, $p = 0.26$). The median hsCRP level in obese group was found statistically significantly higher than in control group (1.97 vs. 0.41, $p < 0.01$). It was found a significant positively correlation between hsCRP and BMI in the obese participants ($r = 0.45$, $p < 0.01$), but not relationship between suPAR and BMI ($r = -0.21$, $p > 0.05$).
Conclusion: Our research did not demonstrate the usefulness of suPAR as an inflammatory biomarker and a predictive value for future atherosclerosis in obese children. Further studies with larger sample size are required to determine whether the suPAR is useful as an inflammatory biomarker in childhood obesity.

P2611 - THIOL DISULPHIDE HOMEOSTASIS AS A NOVEL OXIDATIVE STRESS MARKER IN OBESE CHILDREN

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Objective: Obesity leads to subclinical systemic inflammation and protein/lipid oxidation, and these biochemical changes have also been shown in adult studies in which cardiovascular disease accelerates. Thiol acts as an important antioxidant by forming disulfide bonds to remove reactive oxygen molecules. In this study, we aimed to investigate the usefulness of this novel and sensitive oxidative stress marker, thiol/disulphide homeostasis, in obese children.
Method and Results: Of the total 139 participants, 75 (34 male, 41 female) were in obese group and 64 (27 male, 37 female) were in control group. The median age was 12.0 (5.0-17.0) years old for obese group, and 12.8 (4.7-17.0) years old for control group. Obese children had significantly higher systolic blood pressure, EAT and LV mass than control group ($p < 0.05$). WBC, PLT and hsCRP were higher ($p < 0.01$), but N/L ratio and MCV values were lower in the obese group ($p < 0.001$). Native thiol, total thiol and native thiol/total thiol ratios were lower ($p < 0.05$), disulphide/native thiol and disulphide/total thiol ratios were higher in the obese group compared to the control group ($p < 0.01$). Dynamic disulphide levels were high in the obese group ($p = 0.057$) but not statistically significant.
Conclusion: We found that in obese children, impairment in thiol/disulphide homeostasis, which is indicative of oxidative stress, together with elevation in inflammatory and cardiac markers. We believe that early diagnosis and treatment of patients at risk for complications by evaluating oxidative status in obese children may result in reduced cardiovascular morbidity and mortality in adulthood.

P2857 - LEAN HEALTH STRATEGIES SHOWED IMPROVEMENT IN THE CYCLE OF CARE OF CONGENITAL HEART PROGRAM AN INNOVATIVE TEAMWOK AT FUNDACION CARDIOINFANTIL INSTITUTO DE CARDIOLOGÍA (FCI IC) BOGOTÁ COLOMBIA

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Background: A fundamental part of the standardized objectives of our Congenital Heart Program (CHP) is to provide high quality, timely and accurate diagnosis to improve quality of care. Thus by identifying and implementing proper Lean Management Strategies (LMS) we intend to improve times along the process for better quality of care service.
Objective: to describe the cycle of care of the CHP; to diagram and calculate the time spent at each step along the process by patients and their families. Furthermore we implemented and evaluated LMS according to the identified factors affecting time efficacy.
Methods: This pseudo experimental pilot study evaluated the process and the benefit of LMS in health care services. The time spent in the cycle of care was measured in 180 patients (90 in physician consultation; and 90 in echocardiogram service) since the admission toward discharge. Cycle of care performance (minutes) data were analyzed using analysis of variance (ANOVA) and statistical process control techniques to assess time before and two months after the implementation of LMS. We also used the spaghetti and the Value Stream Mapping (VSM) diagram to investigate time-outs in care. Furthermore we used a DOFA analysis to describe the specialist´s activities.
Results: In terms of health system outcomes, the results of the study showed a scheduling overlap between the specialist consultation and the interventionist procedure. Furthermore the analysis showed patients had to perform many steps ($n = 9$) to attain a medical consultation or a diagnostic examination during the cycle of care requiring 135 minutes. The new intervention reduced the cycle of time to 5 steps spending 45 minutes (a reduction of 90 min).
Conclusions: The lean strategies implemented in the institute of Congenital Heart Disease enhanced the schedules and activities of the service at the time there was an improvement of the level of satisfaction.

P2982 - ETHICS AND RATIONING HOW DOES ONE CHOOSE WHO TO HELP IN A RESOURCE POOR ENVIRONMENT

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With limited operating time patient selection is fraught with ethical difficulties and questions of distributive justice. We practice paediatric cardiac surgery in such a setting, in a well-equipped state-sponsored unit, meeting the needs of only 25% of children with heart disease. Over the past five years 1517 patients have received corrective surgery with a mortality rate of 2.9%. 81% of operations utilized cardiopulmonary bypass. Based on demographic data our potential caseload is over three times greater (900 per year). Access is limited by poor health systems and poverty, so we get away with a first-come first-served system. We feel challenged to consider a more just approach. Options include: • Selection by probable outcomes of quality and duration of life; this is subjective, and impossible to implement fairly. • Basics-only, focusing on volume at the expense of complex lesions; results in poor surgical skills and frustration. • Triage based on urgency and severity; means selecting patients with the worst outcomes. At the centre of the issue is the healing relationship we claim with all children with heart disease, so we are caught between denying the scope of the problem and denying our commitment to all children. When presented with two equally deserving children for a single slot, the only just way is to choose randomly. Hard as it sounds, this excuses us from acting as the arbiter over a child's life, and bypasses the dilemma of who is "most deserving." A first-come first-served system already has a degree of such randomness built into it. The patient-doctor relationship of trust must be preserved. We need to avoid the conundrum of choosing who is more deserving. However because of the relationship, as advocates for our patients we must also pursue innovative approaches and fight to "expand the investment" of authorities.

ADULT CONGENITAL HEART DISEASE

P1015 - BORN WITH A HEART CONDITION THE CLINICAL IMPLICATIONS OF POLYVAGAL THEORY

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Introduction: Since antiquity our hearts have been linked to our feelings in our collective conscious yet, modern medicine has relegated this organ to a functional pump. However, a wide range

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of psychosocial vulnerabilities including anxiety, depression, developmental delay, poorer Quality of Life, infant feeding & oral motor difficulties are associated with being born with a heart condition (Kovacs et al, 2009). To date, these difficulties have been accounted for by secondary factors with recognition & management limited (Morton, 2015; 2014; 2012; 2011).
Methods/Results: Here, I propose Porges' (2011) Poly Vagal Theory (PVT) offers a more holistic account of this condition (Morton, submitted). PVT provides a comprehensive understanding of our nervous system, senses, emotions, social self & behaviours. Porges proposes the nervous system employs a phylogenetic hierarchy of strategies to self regulate & respond to threat, adapting to our environment when we are safe (enabling the 'Social Engagement System') & unsafe (enabling defensive mechanisms), with homeostatic variability shaped during our early years. Since the heart is central to our nervous system congenital cardiac anomalies may compromise our response to stress, emotional regulation & the Social Engagement System. This has implications across the lifespan providing strategies to optimise normal development of social & defensive behaviours & inform therapeutic interventions, explored here drawing on psychological theories & advocacy/personal experience of living with a heart condition from birth.
Based on: Morton, L. (submitted) Born with a heart condition: The Clinical Implications of PVT in Clinical application of PVT around the world (working title) Ed: Porges, S & Dana, D. Norton Professional Books: New York.

P1029 - CORRELATION OF FRAGMENTED QRS COMPLEX WITH RIGHT VENTRICULAR INDICES AFTER REPAIR OF TETRALOGY OF FALLOT

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Background: Post-operative right ventricular (RV) progressive dilatation and dysfunction especially with the development of pulmonary regurgitation (PR) is increasingly recognized after repair of tetralogy of Fallot (TOF).Fragmented QRS (fQRS) complex reflects conduction delay due to myocardial fibrosis is a proposed preliminary simple screening tool for such changes.
Materials and Methods: Thirty patients with repaired TOF were included. All patients had contemporary ECG and CMR. fQRS complex was defined as the presence of >2 R waves or >2 notches in the R/S waves in case of wide QRS complex and the presence of an additional R wave or notched S wave in two contiguous leads in case of narrow QRS complex.
Results: The mean age was 15 ± 3.13 years, time passed since repair 11.5 ± 1.87 years; fQRS was present in 70% of pts. fQRS was correlated to higher RV end-diastolic volume index (RVEDVI) (p <0.001), RV end-systolic volume index (RVESVI) (p <0.001), higher PR fraction (p = 0.004), and lower RV ejection fraction (RVEF) (p = .018). A cutoff point of fQRS in 5 leads showed a sensitivity of 87.5% and specificity of 85.7% for detection of RVEDVI >150 ml/m2, and 83.3% sensitivity, 66.7% specificity in identifying patients with PR fraction more than 45%.Prolonged QRS duration was correlated to higher (RVEDVI) (p = 0.004), (RVESVI) (p = 0.006), with a trend towards lower RVEF (p value = .145) and higher (PR) fraction (p = .175).
Conclusions: fQRS was correlated with higher (RVEDVI), (RVESVI), (PR) fraction, and lower (RVEF), furthermore the presence of 5 fQRS leads is a sensitive and specific predictor of RVEDVI >150 ml/m2, and PR fraction more than 45%.

P1030 - QRS PRODUCT AS A NEW ECG TOOL FOR SCREENING OF PROGRESSIVE RIGHT VENTRICULAR DILATATION AND SEVERE PULMONARY REGURGITATION AFTER REPAIR OF TETRALOGY OF FALLOT

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Background: Prolonged QRS duration and QRS fragmentation were used as a simple ECG tools for screening of progressive right ventricular (RV) dilatation in patients with repaired tetralogy of Fallot (TOF).To the best of our knowledge, this is the first study that correlate the product of both ECG tools with RV indices in repaired TOF.
Materials and Methods: Thirty patients with repaired TOF were included. All patients had contemporary ECG and CMR. Fragmented QRS complex (fQRS) was defined as the presence of >2 R waves or >2 notches in the R/S waves in case of wide QRS complex and the presence of an additional R wave or notched S wave in two contiguous leads in case of narrow QRS complex. The product of (sum fQRS leads × QRS duration in ms) was calculated and named "QRS product". A sum of one was given for patients with no fQRS.
Results: The mean age was 15 ± 3.13 years, time passed since repair 11.5 ± 1.87 years; fQRS was present in 70% of patients. QRS product showed significant linear correlations with RV end-diastolic volume index (RVEDVI) (p value <0.001), RV end-systolic volume index (RVESVI) (p value <0.001), Pulmonary regurgitation (PR) fraction (p value = .006) and significant inverse correlations with RV ejection fraction (RVEF) (p value = .019). The optimum QRS product cutoff point for detection of patients with (RVEDVI) more than 150 ml/m was 710 with a sensitivity of 87.5% and specificity of 85.7%, and 780 for (PR) fraction more than 45% with a sensitivity of 83.3%, and specificity of 83.3%.
Conclusions: QRS product was correlated with higher (RVEDVI), (RVESVI), (PR) fraction, and lower (RVEF), furthermore QRS product of 710 was a sensitive and specific predictor of (RVEDVI) > 150 ml/m2, and QRS product of 780 was a sensitive and specific predictor of (PR) fraction more than 45%.

P1041 - DIFFERENTIAL PULMONARY ARTERY REGURGITATION POST TETRALOGY OF FALLOT REPAIR A CARDIAC MAGNETIC RESONANCE STUDY

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Background/Hypothesis: Pulmonary regurgitation (PR) is an important cause of late morbidity after tetralogy of Fallot (TOF) repair but little is known about some of its underlying mechanisms. This phase-contrast cine magnetic resonance (PC MR) study was performed to evaluate the contribution of each pulmonary artery (PA) to the regurgitant volume and its effects on right ventricular (RV) parameters.
Patients and Methods: Forty patients with significant PR underwent a PC MR after TOF repair.
Results: Forward flow in the right PA (RPA) was significantly higher than in the left artery (LPA) [53.2 ± 15.6 vs 34.8 ± 17.7, p < 0.001] and RPA net flow was also higher than LPA [33.9 ± 10.9 vs 17.9 ± 10.7, p <0.001]. Although LPA regurgitant fraction (RF) was significantly higher than that of RPA [45.2 ± 15.9 vs 36.4 ± 13.4, p: 0.008], its relative contribution to

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main PA regurgitation (MPA-R) was significantly lower [40.9 ± 18.6 vs 51.6 ± 15.9, p: 0.0047]. LPA contribution to MPA-R was significantly higher when TOF repair was done at an age >10 years (51.7 ±17.1 vs 36.6 ± 17.4, p: 0.016). Severe MPA-RF ≥ 40% had significantly higher RV endsystolic volume (RVESV) [94.3 ± 45.9 vs 67.5 ±31.9, p: 0.037] and RV end-systolic volume index (RVESVI) [72.6 ± 30.5 vs 52.5 ± 17.7, p: 0.012] than MPA-RF <40%, but they had similar ejection fraction (RV-EF) [53.8 ±11.2 vs 56.6 ±7.5, p : 0.35].
Conclusions: RPA relative contribution to PR severity was higher than LPA in follow up of patients with severe PR after TOF repair. LPA contribution was more in repairs done after the age of 10 years which may help in future management using focal interventions to reduce the regurgitation. RVESV and RVESVI were significantly associated with severe MPA-RF but not RVEF, thus they can be used in the follow up of these patients.

P1042 - CHANGING FACE AND OUTCOME OF INFECTIVE ENDOCARDITIS AND CONGENITAL HEART DISEASE A SINGLE INSTITUTION STUDY IN EGYPT

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Background/Hypothesis: Infective endocarditis (IE) is still a major source of morbidity and mortality worldwide. Rheumatic valvular disease – which is becoming less prevalent - is still the main risk factor for developing endocarditis in Egypt but CHD is steadily increasing as a risk as they become adults. The purpose of this study was to describe the epidemiology of infective endocarditis in Egypt.
Methods: This is a retrospective descriptive study. Electronic data bases were reviewed. All patients referred to the endocarditis working group of Cairo University Hospitals with a diagnosis of definite/possible infective endocarditis from February 2005 till June 2015 were included.
Results: Patients enrolled were 376, thirty three of them (8.8%) had congenital heart disease with a mean age of 32.5 years. Males were by far more common and were generally referred late. Health care associated endocarditis was present in 18.2%. Prior antibiotic use was documented in 72.7% and culture negative endocarditis was seen in 56.7% of whom 15% were diagnosed by serology and/or tissue culture. Seventy nine% of patients were indicated for surgery which was performed in 69.2%.
Conclusions: CHD is a well-established risk factor for infective endocarditis and complex techniques used for treatment are themselves added risks. Although at higher risk, congenital heart disease patients did not have a worse prognosis. A high incidence of zoonotic infections among the whole group and a long time interval from symptom onset till referral was also noted increasing the rates of morbidity and mortality in this group. More effort should be made to diagnose children with asymptomatic congenital heart disease for better future endocarditis prevention regimens as well as encouraging better oral hygiene at homes and schools.

P1044 - MOBILE HEALTH PROGRAM TO PROMOTE SELF MANAGEMENT IN YOUTHS WITH CONGENITAL HEART DISEASE DESIGN AND DEVELOPMENT OF THE COOL RANDOMIZED CONTROLLED TRIAL

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Background: The transition from family-centered nursing care to self-care for youths with congenital heart disease (CHD) commonly occurs in late adolescence to early adulthood. However, regarding how to manage the complex healthcare needs of these youths undergoing transitional care would be a challenge. Mobile health initiatives may deliver CHD information and provide a platform for promoting their self-management.
Aims: to describe the design and development of the Care and Organize Our Lifestyle (COOL) program for youths of CHD and to highlight the importance of mobile health intervention design that made it possible.
Methods/Design: The COOL program was a 12-month randomized controlled trial comparing two active interventions to a usual-care control group. Participants were 150 patients with mild and moderate CHD aged 15-24 years in pediatric and adult CHD outpatient departments. All participants were offered exercise measurement wristband. Both active interventions were designed based on self-regulation theory and incorporated techniques for behavioral self-management and motivational enhancement. One active intervention was provided COOL passport, a mobile healthcare App. The other was provided health promotion cloud, an interactive platform along with COOL passport. The primary outcome was change in cardiopulmonary exercise test and the International Physical Activity Questionnaire (IPAQ)-Taiwan Show-Card Version score. Cardiac knowledge measured by Leuven knowledge questionnaire for congenital heart diseases was a secondary outcome.
Discussion: The trial is expected that the complete COOL program will be scalable to promote self-management in youths with CHD, with the potential for national-level adoption.

P1054 - THE DUCT DIAGNOSIS IN ICU PRETERME INFANT DIAGNOSTIC TOOL

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Introduction: Patent ductus arteriosus /PDA/ is a large vascular structure with an important role in the foetal circulatory pattern, most often is closed within 72 hrs after birth, but in premature infants closing may be delayed a few days, even weeks, leading to a significant comorbidity. The aim of the study was to correlate the sensitivity of diagnostic tools in PDA detection.
Patients and Methods: A prospective study conducted in 2015-2016, at Neonatal Cardiology Intensive Care Unit, Pediatric Clinic of University Clinical Centre Sarajevo, included 20 premature infants, age 28-34 WG, with PDA. Control group formed 20 premature infants. Correlation of echocardiographic, clinical and phonocardiographic examination was realized in each child.
Results: There was no statistically significant difference between the study and control group in gender, the average gestational age, birth weight, Apgar score, pulse, precordial activity, femoral pulsation ($p > 0.05$). There was a statistically significant difference ($p < 0.05$) between the control and study group compared to auscultatory finding on heart, as well as statistically significant ($p < 0.05$) relationship between physical finding at the heart and the diameter of the ductus. Correlation of phonocardiographic findings, showed statistically significant ($p < 0.05$) differences between the study and control group with respect to the frequency of the

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murmur but without statistically significant ($p > 0.05$) difference in relation to the murmur character.
Conclusion: Color Doppler echocardiography is more sensitive tool in PDA detection in preterm infants in relation to clinical and phonocardiographic findings which, as screening diagnostic modality, contribute to the start of adequate therapy and shorter stay of preterm pts at ICU.

P1058 - COARCTATION LONG TERM FOLLOW UP AND QUALITY OF LIFE PREDICTIVE VALUE OF CLINICAL VARIABLES

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Objective: Long-term sequelae and events after coarctation repair are well described. However, the predictive value of variables from clinical follow-up investigation for late events has rarely investigated.
Methods: All patients, who have participated in the prospective cross-sectional COALA Study in 2000 with a structural clinical investigation including blood pressure measurement and symptom-limited exercise test were contacted for reevaluation of current clinical status, medical treatment, office and ambulatory blood pressure measurement and the health-related quality of life questionnaire SF-36.
Results: From 273 eligible patients, 153 patients responded, 27 patients explicitly denied to participate in the study. Nine patients died during the follow-up time with the age of 46 years (range 30-64 years), 25 patients had an important event (12 procedures at the aortic valve due to bicuspid aortic valve, 8 procedures for recoarctation, 2 endocarditis, 2 cerebral insults,1 endocarditis). Thirty-five patients (30%) showed normal ambulant blood pressure, 56 (48%) were on antihypertensive medication and 25 (22%) had arterial hypertension without receiving medication. Quality of life was good in the fields of physical role and pain. However, patients reported a significant impairment in general health, depending on the age. Arterial hypertension, echocardiographic measurements or exercise capacity from the COALA study were not predictive on functional health status.
Conclusion: Repaired coarctation of the aorta shows fairly low mortality on long-term follow-up. Important events are not only recoarctation, but also procedures at the bicuspide aortic valve. The rate and the severity of arterial hypertension is progressively increasing. However, the predictive value of clinical variables is limited, except the presence of a bicuspid aortic valve for aortic valve procedures.

P1059 - EARLY PULMONARY ARTERIAL HYPERTENSION AFTER CLOSURE OF A VENTRICULAR OR COMPLETE ATRIOVENTRICULAR SEPTAL DEFECT BEYOND 6 MONTHS OF AGE

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Background: Pulmonary arterial hypertension (PAH) is virtually absent after closure of ventricular septal defect (VSD) in the first six months of life. However the prevalence of PAH in patients, who underwent VSD closure later, is not clear. The aim of this study was to analyse the prevalence of PAH after a successful VSD closure after the age of 6 months and whether there are risk factors for developing PAH.
Methods: Echocardiographic and right heart catheter data of patients with VSD or complete atrioventricular septal defect, who underwent VSD closure after the age of 6 months in our institution between 01/2005 and 06/2014, were retrospectively analyzed. PAH was defined as mean pulmonary arterial pressure (mPAP) of ≥25 mmHg or tricuspid regurgitation jet velocity of ≥ 3.5 m/s.
Results: In 228 patients (median age at shunt closure 4.0 years, range 0.5-69) and 174 complete follow-up data (median follow-up 3.7 years, range 0.5-39.4), 9 patients needed pulmonary vasodilator therapy after shunt closure, 4 of them temporarily for up to 79 months. Three patients are still on vasodilator treatment 1, 2.6 and 6 years after surgery, other two were lost to follow-up. Another 6 patients with preoperatively borderline hemodynamics due to elevated mPAP and pulmonary vascular resistance, recovered well without signs of postoperative PAH.
Conclusion: With the current practice for safe late VSD closure, PAH is very rare at least in the first years of follow-up. In most patients with perioperative PAH, this condition appears to be transient and shows good response on pulmonary vasodilator treatment.

P1061 - MANAGEMENT OF TACHYARRHYTHMIAS IN PATIENTS WITH ADULT CONGENITAL HEART DISEASE AFTER CARDIAC SURGERY

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Background: Tachyarrhythmia is a common late complication in patients with adult congenital heart disease (ACHD) after surgery.
Purpose: The aim of this retrospective study is to investigate clinical results of interventional therapies for various tachyarrhythmias complicated with ACHD.
Methods and Results: 180 patients with the median age of 33 years old were investigated. 160 patients had supraventricular tachycardia (SVT) and 20 patients had ventricular tachycardia (VT). The previous surgery with SVT was APC-Fontan in 70 patients, TCPC-Fontan in 22, ASO (atrial switch operation) in 12 and biventricular repair (BVR) in 56. We performed catheter ablation (CA) in 139 patients with SVT. Acute success rate of CA was 66% (34/51) in APC, 70% (14/20) in TCPC, 91% (11/12) in ASO and 85% (48/56) in BVR. Recurrence rate after CA was 26% (9/34), 36% (5/14), 27% (3/11) and 13% (6/48), respectively. Antiarrhythmic drugs was also used in 80% (56/70), 68% (15/22), 50% (6/12) and 48% (27/56), respectively. A pacemaker was implanted to control SVT in 31% (22/70), 22% (5/22), 41% (5/12) and 26% (15/56), respectively. The previous surgery with VT was RVOT reconstruction in 10, Rastelli in 7, APC-Fontan in 1, ASO in 1, AVR in 1. CA was performed in 40% (8/20), acute success rate was 50% (4/8). ICDs or CRT-Ds were implanted in 85% (17/20) and drugs were used in 95% (19/20).
Conclusion: CA can be a curative approach especially for SVT of postoperative ACHD. However, hybrid therapy with antiarrhythmic drugs or cardiac implantable electric devices may be required in the selected population.

P1076 - FACTORS INFLUENCING LOSS TO FOLLOW UP AMONG PATIENTS WITH CONGENITAL HEART DISEASE IN MALTA PRIOR TO THE INTRODUCTION OF FORMAL TRANSITION – UNDERSTANDING THE AVOIDABLE

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Background: While most congenital heart disease (CHD) is successfully managed in childhood, lifelong follow-up is required to detect and manage significant recurrent or residual lesions and arrhythmias. Loss to follow-up of patients is a set-back to this surveillance process. The aim of this retrospective study was to investigate factors influencing loss to follow-up among Maltese adult patients with CHD of moderate/great complexity prior to institution of formal transition.
Materials and methods: 187 patients - tetralogy of Fallot 70, coarctation/interrupted aortic arch 56, atrioventricular septal defect 34, Fontan-type palliation 13, transposition of great arteries 14 - aged 16 years or over by end 2013 were included. Clinical details and follow-up records were obtained from hospital notes. Ordinal logistic regression was applied to investigate the potential influence of (a) gender (b) CHD complexity (c) dedicated paediatric cardiology input in childhood (d) number of cardiac surgeries/interventions and (e) long-term cardiac medications, on loss to follow-up.
Results: 41/187 patients (21.9%; 27 males) were lost to follow-up. Limited input by paediatric cardiologists in childhood, a smaller number of surgeries/interventions and lack of long-term cardiac medications were all identified as independent predictors for loss to follow-up in our study population. CHD lesion complexity of itself was not significantly associated with loss to follow-up (Table).
Conclusions: This study identifies CHD patient characteristics that increase the chances of loss to long-term specialist follow-up. Patients with such characteristics warrant closer attention, particularly at time of transfer to adult cardiology care. In particular, it underlines the importance of an infrastructure that provides input by paediatric cardiologists in the care of these patients from an early stage as a means of ensuring appropriate specialist follow-up throughout life.

Table. Impact of studied factors on loss to follow-up.

Factor	OR	95% CI lower, upper	p value
Male vs. female gender	2.12	0.80, 5.65	0.13
Moderate vs. great complexity	1.60	0.32, 8.03	0.57
Limited vs. structured paediatric cardiology input in childhood	5.08	1.77, 14.63	0.003*
Up to 1 vs. more than 1 cardiac surgery/intervention	3.34	1.09, 10.26	0.04*
No vs. on long-term cardiac medication	7.34	1.74, 31.02	0.007*

* Statistically significant association (statistical significance defined as $p < 0.05$)

P1083 - ECHOCARDIOGRAPHIC ANALYSIS USING NEW TECHNICS TO EVALUATE VENTRICULAR FUNCTION RESULTING FROM CHRONIC PULMONARY REGURGITATION IN PATIENTS WITH FALLOT

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Methods: 73 patients with 19 ± 4 years (y). Average age at surgical repair: 2.7 ± 1 y. Evaluated with Tissue Doppler (TDI), Tissue track, TEI Index, Ejection Fraction, 2 D Speckle Tracking (STD), basal and mid Strain in free Wall of both ventricles; magnetic resonance was done when patients registered increase in ventricule dysfunction or when the volume was >120 ml/m^2 by echocardiography. Patients were divided into 3 groups: 1) right ventricle (RV) <100 ml/m^2, 2) RV more than 120 ml/m^2 but less than 150 ml/m^2, 3) RV > 150 ml/m^2.
Results: In the first group with RV < 100 ml/m2 there were no significat changes, except for systolic velocity of TDI which decreases in mid RV , no changes in regional strain and strain rate. Theses patients have no important repercussion in RV 2DSTD; 88% of the cases recovered normal function after valve repair. The second group evidenced RV and LV dysfunction in TDI (inversion of diastolic wave), global Strain decrease, and regional basal and mid Strain were opponents to normal in RV and basal LV. The third group had severe dysfunction in LV and RV 2DSTD, and opponent curves in regional basal and mid strain in both ventricles.
Conclusions: the regional myocardial deformation can be evaluated from echocardiography-derived speckle-tracking analysis to measure myocardial Strain and provides direct information of the contractile performance of the right and left ventricle. The new eco technology can identify early LV repercussion and early PVR may be the new criteria to prevent LV irreversible damage when the LV dysfunction increases.

Table. Fallot global deformation 2DSTD

GLOBAL 2D STD	GROUP 1		GROUP II		GROUP III	
N patients	X	DS	X	DS	X	DS
body surf.	18		23		32	
VFD RV ml/m2	1,35	0,31	1,11	0,42	1,5	0,26
	68	18	114,33	4,51	151,82	20,43
RV						
2D STD 5c RV %	-27,4	2,03	-17	2,83	-14,43	5,26
4c RV %	-19,6	3,02	-19	3,04	-15,28	4,17
2c RV %	-22,8	2,02	-18	0	-15,29	4,88
AVG %	-23,3	2,02	-17,8	2,55	-14,68	3,61
LV						
2D STD 5c LV %	-20,1	1,05	-17,67	0,58	-15,29	4,88
4c LV %	-25,5	1,02	-17	2,65	-14,64	2,65
2c LV%	-18	2	-17	2,6	-11,82	4,32
AVG %	-19,7	1,07	-15,5	3	-14,52	3,16

P1102 - ARRHYTHMIA BURDEN AND RELATED OUTCOMES IN EISENMENGER SYNDROME

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Background: Patients with Eisenmenger syndrome have a shorter life-span than the general population. A significant proportion develop arrhythmia and some, sudden death.

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Objective: The aims of this study was to characterize the frequency, type and effects of arrhythmias in a moderately large cohort of adult patients with Eisenmenger's syndrome and to identify potential risk factors for arrhythmia.
Methods: This retrospective study included patients aged ≥ 18 years of age with Eisenmenger's syndrome from Southampton University Hospital; Japanese Adult Congenital Heart group and Royal Victoria Hospital. Information was collected from chart review and arrhythmias were noted from electrocardiograms and Holter studies.
Results: A total of 167 patients were included with a mean age of 37 ± 10 years (range: 18–63 years) with the majority in NYHA functional class II or III. Twenty eight patients (17%) had a documented arrhythmia: paroxysmal supraventricular tachycardia (8 patients, 29%), atrial fibrillation (6 patients, 21%), atrial fibrillation and flutter (2 patients, 7%), non-sustained ventricular tachycardia (6 patients, 21%) and sustained ventricular tachycardia (6 patients, 21%). Among the entire study group, 25 patients (15%) were currently on anti-arrhythmic therapy and 77 patients (49%) were on disease targeting therapy for pulmonary hypertension. Downs syndrome was present in 76 patients (46%). Patients with arrhythmia were older [p = 0.01] and were more likely to have atrioventricular valvar regurgitation [Odds ratio: 4.33]. Arrhythmias were found to independently predict all-cause mortality [p = 0.004] and sudden death in bivariate analysis [p = 0.03]. Disease targeting therapy was a protective factor against all-cause mortality in logistic regression analysis [Odds ratio: 0.31], while anti-arrhythmics predicted sudden death [Odds ratio: 6.24].
Conclusion: Arrhythmias are common among patients with Eisenmenger syndrome, especially among those with atrioventricular valve regurgitation. Arrhythmias may predict sudden death, the risk of which might not be reduced by anti-arrhythmics.

P1136 - TRANSITION CONGENITAL HEART DISEASE SERVICE

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Introduction: Transfer from paediatric to adult services is a stressful time for the adolescent and their parents/carers. Becoming independent is a giant step in their lives. Our service aims to educate, inform and empower, preparing them for the adult world with responsibilities and decision making.
Background: Survival rates are ever increasing for young adults with Congenital Heart Disease into adult life. This is mainly due to improvements in medical and surgical management and in more recent times, the support of the nurse specialist. Research suggests that many of these patients are inadequately prepared for their future care. There is lack of on-going education to help prepare these young people to manage their health care needs into adulthood; many will require lifelong cardiac surveillance. A very significant issue for this population are those lost to follow-up and unfortunately present when a significant cardiac problem arises which could have been prevented, through regular check-ups. The educational process needs to be structured, ensuring uninterrupted health care, patient centred, age and developmentally appropriate, flexible and comprehensive aiming improvements in health status, quality of life and survival outcomes. Transition starts at the age of 12 and finishes at 25.
Method: The service offers nurse-led clinics and nurse led day cases where the adolescent can attend with or without their parents/carers. Telephone clinic consultations are offered as well as support via email.
Results: Between 14/01/2016 and 30/11/2016 413 adolescents were seen in clinic. The feedback has been very good demonstrating support for our unique service.
Conclusions: A co-ordinated transfer with a summary document is an important step in a successful transition process. Patients seen expressed satisfaction defining the service as effective, professional and supportive of their autonomy for taking the lead for their own care.

P1146 - COMPARISON BETWEEN 2 DIMENTIONAL ECHOCARDIOGRAM AND CARDIAC MAGNETIC RESONANCE IN ASSESSING RIGHT VENTRICULAR SIZE AND FUNCTION IN PATIENTS WITH REPAIRED TETRALOGY OF FALLOT

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Background: Right ventricular (RV) enlargement and dysfunction are common sequela from significant pulmonary regurgitation (PR) following tetralogy of Fallot total correction (TFTC). Accurate assessment of RV size and function is important during follow-up and consideration of pulmonary valve replacement (PVR). Limited studies compared 2-dimentional echocardiogram (2DE) with cardiac magnetic resonance (CMR) in the assessment of RV size and function in adult TFTC patients. We sought to compare measurements of RV size and function by 2DE with CMR.
Methods: We studied 88 selected patient with moderate-severe PR (43 male; median age = 30, range = 17-73y) after TFTC. 2DE measurements included RV basal, major and minor at end-diastole, annular plane systolic excursion (TAPSE) and tricuspid annulus tissue Doppler velocity (RV S'). On CMR, the RV was reconstructed from manually traced borders to calculate RV volumes and ejection fraction (RVEF). Correlation between 2DE and CMR parameters was analysed using linear regression. Indications for PVR based on CMR derived RV volumes and RVEF (i.e. RVEDVI > 150 ml/m2, RVESVI > 80 ml/m2 or RVEF < 47%) were used as outcomes in univariate and multivariate Cox regression analyses.
Results: 2DE RV dimensions correlated weakly to RV volumes (r = 0.38-0.53; p < 0.01). There were no correlations between TAPSE and RV S' with RVEF (r = -0.06; p = 0.57 and r = 0.16; p = 0.12 respectively). Among RV measurements, only RV major was independently associated with dilated RV on CMR in the univariate analysis but rendered non-significant after adjusting for age, BSA and gender. TAPSE and RV S' were both independently associated with RV dysfunction in the univariate analysis but rendered non-significant after adjusting for age, BSA and gender (Table 1).
Conclusion: 2DE quantification of RV size correlated weakly with RV volumes and TAPSE and RV S' did not correlate with RVEF in adult patients with TFTC. Quantification of RV size and function by 2DE was not associated with significant RV dilatation and dysfunction, especially those requiring PVR.

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Table 1. Relationship between 2DE and CMR parameters in patients with TFTC. (n = 88)

	Univariate analysis			Multivariate analysis		
	OR	95% CI	P-value	Adjusted OR	95% CI	P-value
Relationship between RV dimensions and RVEDVi >150 ml/m^{2}or RVESVi > 80 ml/m^2						
RV basal	1.02	0.99-1.06	0.27	1.04	0.98-1.11	0.19
RV basal >41 mm	1.14	0.58-2.25	0.71	1.45	0.63-3.34	0.39
RV major	1.03	1.00-1.05	0.04	0.98	0.94-1.02	0.30
RV major >83 mm	1.81	0.99-3.30	0.05	1.02	0.47-2.22	0.95
RV minor	1.01	0.98-1.05	0.51	0.99	0.93-1.05	0.66
RV minor >35 mm	1.37	0.75-2.51	0.31	1.00	0.48-2.10	0.99
Age (years)	0.78	0.71-0.85	<0.01	0.78	0.72-0.85	<0.01
BSA (m^2)	1.44	0.47-4.37	0.52	0.45	0.08-2.38	0.35
Male gender	0.46	0.25-0.84	0.01	0.62	0.26-1.48	0.28
Relationship between TAPSE and RV S' and RVEF <47%						
TAPSE on 2DE	0.84	0.76-0.94	<0.01	0.90	0.79-1.03	0.13
TAPSE <1.7 cm	2.59	1.30-5.12	<0.01	1.43	0.46-4.43	0.54
RV S'	0.92	0.79-1.07	0.27	0.93	0.75-1.14	0.48
RV S' <10 cm/s	1.43	0.74-2.77	0.28	1.62	0.58-4.50	0.36
Age (years)	0.85	0.80-0.91	<0.01	0.86	0.80-0.92	<0.01
BSA(m^2)	1.73	0.49-6.10	0.40	0.55	0.09-3.40	0.52
Male gender	0.35	0.17-0.72	<0.01	0.45	0.17-1.20	0.11

P1150 - NEUROLOGICAL OUTCOME OF CONGENITAL HEART DISEASE WITH VERY LOW BIRTH WEIGHT INFANTS

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Objectives: The purpose of the study was to elucidate the neurological outcome in children with congenital heart disease (CHD) born with very low birth weight (VLBW) in Japan.
Methods: A retrospective multicenter cohort study enrolled patients with CHD with VLBWI born from January 2000 to December 2006. Twelve neonatal intensive care unit (NICU) participated in this study. Neurodevelopmental assessment using the Kyoto Scale of Psychological Development (KSPD) was performed in children survived until 3-6years old. We reviewed demographic and KSPD dates of these VLBW infants.
Results: Forty-five patients were enrolled in this study. The median birth weight was 1148 g (range504-1477g) and median gestational week was 31weeks (range 23-39weeks). The most common lesions were ventricular septal defect (VSD) (44.4%), tetralogy of Fallot (15.6%), pulmonary valve stenosis (8.9%). The genetic syndromes and extra-cardiac malformations were seen in 20.0%, 31.1%, respectively. Surgical and/or catheter interventions were performed in 21 infants (46.7%) and 17 infants (81.0%) were achieved to the final operation. Overall Developmental Quotient (DQ) was 80.5 (range 19-105). Language-Social (L-S) domain DQ was 81 (range 16-114). Postural-Motor (P-M) domain DQ was 76.5 (range 12-124). Cognitive-Adaptive (C-A) domain DQ was 83(range 14-117).The score of treated patients was poorer than untreated patients in all parameters (overall DQ 62.0 ± 27.8 vs. 84.1 ± 20.0, p < 0.01, L-S DQ 60.3 ± 28.5 vs. 83.1 ± 20.0, P < 0.01, P-M DQ 60.7 ± 31.6 vs. 86.8 ± 24.7,p < 0.01, C-A DQ 65.9 ± 29.2 vs. 84.7 ± 21.1, P < 0.05) respectively.
Conclusions: Neurological functions of VLBWI with CHD survived until childhood are lower limit of reference. Surgical intervention may affect the neurological outcome of VLBWI with CHD.

P1174 - VITAMIN D STATUS AND BONE MASS DENSITY IN ADOLESCENTS WITH FONTAN CIRCULATION

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Background: Although patients with a Fontan circulation are at risk of malnutrition and growth delay, the implications on vitamin D metabolism and bone health have barely been investigated.
Materials and Methods: We prospectively measured serum levels of 25-OH-vitamin D in a nationwide cohort of Fontan patients aged 16 to 18 years old. We also measured bone mass density by narrow fan-beam dual energy x-ray absorptiometry (DXA) scan. The DXA results were compared to age and gender matched reference data and expressed as Z-scores.
Results: Seventeen consecutive patients were recruited from our pre-transition national Fontan clinic. All 17 patients had vitamin D levels <75.0 nmol/l, which has been suggested as a lower limit in patients with chronic illness. Thirteen patients (76%) had vitamin D insufficiency with levels <50.0 nmol/L, 9 (53%) patients had deficiency with values <37.5 nmol/L and two patients (12%) had severe vitamin D deficiency with non-measurable values <12.5 nmol/L. The DXA showed abnormally low z-scores of -1.7 ± 0.9 (mean ± SD) (p < 0.001) for columna and -0.8 ± 1.1 (p = 0.009) for total body measurement respectively. There was no correlation between DXA result and vitamin D level.
Conclusion: Adolescent Fontan patients have a high prevalence of vitamin D deficiency and low bone mass density, however, without these two being correlated. Whether the vitamin D deficiency is due to limitations in sun exposure and dietary intake of vitamin D, or if vitamin D deficiency and insufficient bone mass density are differently related to the Fontan circulation is unknown. Future studies should investigate pathogenesis of both vitamin D deficiency and low bone mass density, as well as determine fracture implications and identify interventions.

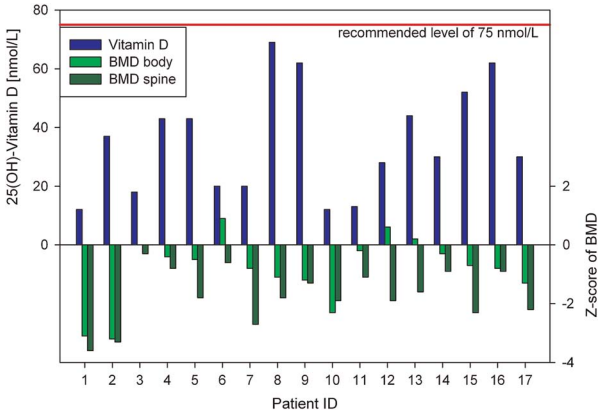

Figure 1.

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P1178 - ACOUSTIC RADIATION FORCE IMAGING OF THE FONTAN LIVER – ADDING TO THE DIAGNOSTIC ARMAMENTARIUM

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Background/Hypothesis: Fontan associated liver disease (FALD) is thought to result from congestion, as a consequence of time post surgery. Biochemistry and imaging do not follow the same rules as other liver injury with most imaging modalities resembling cirrhosis. Routine assessment of adults with a Fontan circulation using biochemistry and Acoustic Radiation Force Imaging (ARFI) to ascertain transient elastography (TE) was undertaken. In other liver diseases, an ARFI of >1.80 m/s has been validated with a high likelihood of cirrhosis.
Materials and Methods: ARFI was performed, by 2 experienced sonographers, in 33 adults (female = 20; median age 28 (22–32.5) years) with a Fontan circulation (atriopulmonary n = 16, lateral tunnel n = 2, extracardiac n = 15). Nodularity on ultrasound (n = 20) identified a subgroup with a higher ARFI score: 2.20 m/s (1.86–2.77) versus 1.87 m/s (1.66–2.13) (2p = 0.039)
Conclusions: There is little published on ARFI in FALD, and our results are higher than reported in other liver disease, with less technically adequate values. The significant difference in time post Fontan completion in our subgroups did not result in differences between ARFI or MELD-XI scores. However, nodularity on US was associated with a higher ARFI score, suggesting that ARFI may have a role in further delineating advanced liver disease. Its accuracy in liver fibrosis short of established cirrhosis, will require further assessment.

Table. Results.

Variable	AP Fontan (n = 16)	LT + ECC Fontan (n = 17)	2p
Age (years)	32 (29–36.3)	22 (20–28)	< 0.001
Years post Fontan	27 (25–31)	16 (13.5–19.5)	< 0.001
ARFI (m/s)	2.11 (1.78–2.57)	2.03 (1.68–2.37)	ns
Technically adequate ARFI readings (n, %)*	14 (88%)	12 (71%)	ns
MELD-XI ^	9.25 (4.60–11.85)	6.72 (4.57–10.27)	ns

Median (IQR); *IQR/median < 0.3; ^MELD-XI = prognostic indicator in liver disease

P1189 - SURGICAL MANAGEMENT OF CONGENITAL HEART DEFECTS BEYOND THE AGE OF 60

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Introduction: The population with congenital heart disease is increasing and ageing. Aim was to assess the results of surgical management of congenital heart diseases beyond the age of 60 years.
Method: Between 2013 and 2016, 112 adults were operated (50 younger, and 62 older than forty years, including 19 patients aged 60-75 years). The numbers of procedures were as follows (in brackets the number of operations in the three age groups): Ross surgery 7 (4, 3, 0); aortic valve replacement 26 (13, 4, 9); subaortic membrane resection 1 (0, 0, 1); Bentall/ascending aortic plasty 18 (7, 8, 3); myectomy with or without mitral valve replacement in left ventricular outflow obstruction 10 (1, 6, 3); aortic coarctation 1 (1, 0, 0); ligation of ductus arteriosus 2 (1, 1, 0); reconstruction of right ventricular outflow tract with biological valve 7 (2, 4, 1); homograft 8 (6, 2, 0); BioValsalva graft 1 (0, 1, 0); primary reconstruction of complete atrioventricular septum defect 4 (2, 2, 0); valve replacement 3 (2, 1, 0); ventricular septum defect 10 (4, 6, 0); atrial septum defect closure 8 (5, 3, 0); total cavopulmonary anastomosis 1 (1, 0, 0); valve replacement in congenital transposition of great arteries 1 (0, 1, 0), Ebstein operation with valve plasty 2 (0, 1, 1); valve replacement 2 (1, 0, 1).
Results: There was no operative mortality, while early mortality occurred in one patient with total cavopulmonary anastomosis due to multiorgan failure.
Conclusions: Congenital heart defects can be operated beyond the age of 60 years with good results in a tertiary heart centre having great experience in the management of congenital and acquired heart disease.

P1193 - MIDTERM OUTCOME OF IMPLANTABLE CARDIOVERTER DEFIBRILLATOR IN REPAIRED TETRALOGY OF FALLOT INFLUENCE OF LONG QT GENES MUTATION POLYMORPHISMS

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Background: Ventricular tachycardia (VT) is a not rare late complication in repaired tetralogy of Fallot (TOF) patients, and implantable Cardioverter Defibrillator (ICD) implantation is indicated in these patients to prevent late cardiac death. Here, using a large repaired TOF cohort, we analyzed the midterm result after ICD implantation and the influence of the long QT gene mutations/polymorphisms.
Method: From 2003 to 2014, all patients of repaired TOF with VT receiving ICD implantation were enrolled. The charts were reviewed and data of ICD was retrieved. We performed mutation analysis of the three common long QT genes (KVLQT1, hERG, SCN5A) through direct sequencing. Patients of repaired TOF without VT served as control group.
Results: A total of 12 patients received (male/female 8/4) ICD implantation. Mean age of TOF repair was 4.4 + -4.5 years, mean age of VT was 25 + -11.1 years, and mean age of ICD implantation was 27.1 + -11.5 years. After mean 5.4 years follow-up, 9 (75%) had at least one episode of nonsustained/sustained VT, 6 (50%) had at least one appropriate shock and 6 (50%) had at least one inappropriate shock. Causes of inappropriate shock include sinus tachycardia in 4, T wave over-sensing in 1, and lead dysfunction in 1. For 6 patients with appropriate shock, 3 attacks were related to drug taper. Use of beta blocker can decrease both appropriate shock and inappropriate shock rate. When compared with 121 control patients, we found those with ICD had higher frequency of long QT genes mutation/polymorphisms (10/12 vs. 54/121, p = 0.014), especially for hERG and SCN5A genes. However, appropriate shock rate has no association with long QT genes mutation/polymorphisms.
Conclusion: The midterm result of ICD implantation in repaired TOF patients showed the appropriate shock rate is high but also inappropriate shock rate. Long QT gene mutations/polymorphisms are associated with necessity of ICD in rTOF patients.

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P1195 - EXPERIENCES OF CHILDREN AND ADOLESCENTS LIVING WITH CONGENITAL HEART DISEASE A SYSTEMATIC REVIEW OF QUALITATIVE STUDIES

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Background/Hypothesis: Congenital heart disease (CHD) is the most common birth defect and can be associated with limitations in physical capacity, neurocognitive impairments, and a predisposition to sudden death. Children with CHD may require ongoing and complex medical intervention, and report psychosocial and lifestyle consequences. We aimed to describe the experiences and perspectives of children and adolescents with CHD.
Materials and Methods: MEDLINE, PsycINFO, CINAHL and Embase were searched in August 2016. Qualitative studies on the perspectives of children aged up to 21 years with CHD were included. Data was analyzed using thematic synthesis.
Results: We included 44 studies from 11 countries reporting the experiences of 995 children and adolescents with CHD. 6 themes were identified: disrupting normality (denying the diagnosis, oscillating between sickness and health, destabilizing the family dynamic); powerlessness in deteriorating health (preoccupation with impending mortality, vulnerability to catastrophic complications, exhaustion from medical testing); enduring medical ordeals (traumatized by invasive procedures, disappointed by treatment failure, displaced by transition, valuing empathy and continuity in care, overcoming uncertainty with information); warring with the body (losing stamina, distressing inability to participate in sport, distorted body image, testing the limits); hampering potential and goals (feeling disabled, unfair judgment and exclusion, incapacity to achieve academically, limiting attainment and maintenance of life milestones); and establishing own pace (demarcating disease from life, determination to survive, taking limitations in their stride, embracing the positives, finding personal enrichment, relying on social/spiritual support).
Conclusions: Children with CHD encounter lifestyle restrictions, fear of invasive procedures, and concerns about body image and discrimination. They endure uncertainties due to the unpredictable progression of disease and a risk of premature death. Access to psychosocial services addressing the distress of medical procedures and community-based support programs may strengthen their coping capacity. This may ultimately lead to improved satisfaction and outcomes in this vulnerable population.

P1205 - CASE REPORT SURGERY ON LUTEMBACHER SYNDROME WITH DEXTROCARDIA AND SITUS INVERSUS

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Background: Lutembacher Syndrome is an uncommon cardiac anomaly characterised by a combination of congenital ostium secundum atrial septal defect and acquired mitral stenosis. Its presence in dextrocardia with situs inversus is an even more unique condition where merely a few reports available. Correcting this unique anomaly thus becomes a challenge for any surgeons. Rheumatic disease is the dominant aetiology of mitral stenosis. Clinical manifestations usually involve dyspnea on exertion due to right ventricular overload leading to right heart failure.
Case Report: A forty-one-year-old male presented with complaints of dyspnea on exertion and palpitation since the last two years. Clinical examination, chest x-ray, echocardiography and catheterisation elicited the diagnosis of Lutembacher syndrome with dextrocardia and situs inversus. The echocardiography showed mitral stenosis insufficiency and an ostium secundum atrial septal defect. The patient was subsequently scheduled for corrective surgery, during which the pericardium adhered firmly onto the heart, indicating a preceding pericarditis. The surgeon substituted the mitral valve with a mechanical replacement and sealed the atrial septal defect with a pericardial patch.
Result: The surgery was a success with no ensuing complications. An echocardiographic evaluation shows a satisfactory result with minimal central valve leakage.
Conclusion: Lutembacher syndrome with dextrocardia and situs inversus is a highly rare entity which presents a challenge in its surgical management. The surgeon corrected this cardiac anomaly with the versatility and excellent technique, resulting in a favourable outcome.

P1208 - A CASE REPORT OF TRUNCUS ARTERIOSUS WITH INTERRUPTED AORTIC ARCH FROM NEWBORN TO ADULT AGE

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We present a case report of a long term follow up results after arterial trunk with type B interrupted aortic arch repair. The prevalence of common arterial trunk is only 1 percent of all CHD. The combination of these diseases is even rarer. The patient was admitted with cardiogenic shock in the newborn age. Immediate surgical approach was reconstruction of aorta and pulmonary artery (PA) banding. Next surgery was done after three more years – the closure of VSD and PA reconstruction with homotransplantat. 13 years later due to progression of PA stenosis and right ventricular failure PA conduit was replaced by percutaneous approach. A 22mm Melody valve was implanted into presented RV outflow tract and 17 mm stent was placed in the right pulmonary artery due to severe stenosis. 20 years after repair patient remains with minimal heart failure symptoms in NYHA I-II class. The positive dynamics of right ventricle systolic function was observed after Melody valve implantation by heart MRI. RV ejection fraction increased from 30 to 43 percent. RV decreased in size without signs of pressure overload. Minimal RV outflow tract dilatation with hypertrophy is still visualized by ultrasound. Both ventricles are in good systolic function. Minimal PA stenosis with minimal PA regurgitation remains. A trivial aortic annulus and root dilatation with minimal aortic regurgitation persists without negative dynamics in past few years. The patient is well socially adapted she evaluates her life quality as great. Prognosis of this particular case: a PA conduit replacement in few more years and real possibility for the aortic valve replacement in late future. Treatment of complex CHD is a challenge for patient, his family and doctors. Correct diagnostic approaches, successful surgical and invasive treatment preserves good systolic function of both ventricles which determines clinical status and life quality of the patient.

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P1210 - PREVALENCE OF BRADYCARDIA ATRIOVENTRICULAR BLOCK AND BUNDLE BRANCH BLOCK IN PATIENTS WITH ATRIAL SEPTAL DEFECT

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Background: Atrial septal defect (ASD) is the second most common type of congenital heart disease. Lately it has been revealed that patients with ASD have increased risk of stroke, pneumonia and atrial fibrillation, emphasizing the need of reconsidering a long-term medical follow up. In order to construct a better follow up procedure for ASD patients, we aim to elucidate whether ASD patients have an increased risk of conduction abnormalities compared to the background population, as consequences of these conditions can be prevented, e.g. with an implantable pacemaker.
Methods: We included 146 patients in this retrospective cohort study where data was collected by reviewing medical records and analysing electrocardiograms (ECGs) for the following outcomes; atrioventricular block (AVB), bradycardia, complete right bundle branch block (CRBBB), incomplete right bundle branch block (IRBBB), left anterior fascicular block (LAH), left posterior fascicular block (LPH), pacemaker and mortality.
Results: Most frequent findings among ASD patients were IRBBB (50%), LAH (5.5%) and bradycardia (4.8%). In total 7 patients (4.8%) had received a pacemaker. Average age at implantation was 20 years. Less common conditions were: 1-AVB (2.1%), 2-AVB (2.7%), 3-AVB (1.4%), CRBBB (3.4%) and LPH (2.1%). Furthermore, we found a low mortality (4.3%).
Conclusion: To date no other European study has examined the prevalence of conduction abnormalities in ASD patients, and our study contributes to an incremental advance of knowledge concerning possible long-term effects of ASD. We found a higher prevalence of AVB, IRBBB and CRBBB compared to a background population, and we discovered that different ages contain different types of conduction problems. The latter may indicate that IRBBB may regress or evolve into a CRBBB with time. Furthermore, we found that patients with ASD are prone to receive pacemaker in a very young age compared to what is expected in a general population of Denmark.

P1215 - INSPIRATORY MUSCLE TRAINING IMPROVES INSPIRATORY MUSCLE STRENGTH RESTING CARDIAC OUTPUT AND THE VENTILATORY EFFICIENCY OF EXERCISE IN PATIENTS WITH A FONTAN CIRCULATION

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Background: Patients with a Fontan circulation have reduced exercise capacity and respiratory muscle weakness. Inspiratory muscle training (IMT) improves exercise capacity and quality of life in adults with heart failure, who have a similar physiology to Fontan patients. We assessed if IMT improves inspiratory muscle strength, measured by maximal inspiratory pressure (MIP), and objective measures of exercise capacity, specifically ventilatory efficiency as measured by VE/VCO2 slope, in adolescents with a Fontan circulation.
Methods: Twenty-three adolescents (aged 16 ± 2 years) with a Fontan circulation underwent 6 weeks of IMT for 30 minutes daily. Respiratory muscle strength (MIP and maximal expiratory pressure, MEP), lung function and exercise capacity (cardio-pulmonary exercise testing) were assessed. Fourteen of the 23 subjects also underwent an exercise cardiac magnetic resonance imaging (ex CMRI) to examine the effects of IMT on systemic and pulmonary blood flow.
Results: Baseline MIP was 69 ± 22 cmH2O, higher in males than females (75 vs 62cmH2O, $p = 0.02$). Six weeks of IMT improved MIP by 36 ± 24cmH2O ($61\% \pm 46\%$). There was an associated improvement in VE/VCO2 (34.2 vs 32.2, $p = 0.04$). There was also improvement in resting cardiac output (from 4.2 ± 1.2 to 4.5 ± 1.0 L/min, $p = 0.03$) and ejection fraction (from 50.1 ± 4.3 to 52.8 ± 6.1, $p = 0.03$) in those who underwent ex CMRI.
Conclusions: Six weeks of IMT improves inspiratory muscle strength, VE/VCO2 slope, and resting cardiac output and ejection fraction, in young Fontan patients. IMT may be a simple beneficial addition to the current management of Fontan patients, potentially improving exercise capacity, morbidity and mortality.

P1224 - SUCCESSFUL TRANSFER OF YOUNG ADULTS FROM PAEDIATRIC TO ADULT CONGENITAL HEART DISEASE SERVICES; REDUCING LOSS TO FOLLOW UP

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Background: Loss to follow-up at transfer from paediatric to adult congenital cardiology services is a recognized challenge in maintaining high quality care of young people with congenital heart disease (CHD). International studies report 7 - 76% of adolescents with CHD are either lost to follow-up or experience lapses in care on transfer to adult congenital heart disease (ACHD) services. This is associated with increased morbidity and mortality. Having a coordinated process with dedicated staffing and resources overseeing transition is the key to effective transfer. This study was performed to assess successful transfer from paediatric cardiology to adult congenital cardiology services within the public health sector in Western Australia, with the introduction of a structured transition programme and a clinical nurse specialist (CNS) who works within both services.
Method: We reviewed all patients aged 16 years and over at time of last paediatric cardiology outpatient clinic review over a 5 year period (01/01/2011-31/12/2015). Included were all patients planned for transfer from paediatric to ACHD services. Patients planned for transfer to other adult cardiology services and ACHD patients receiving services from regional cardiology services were excluded from the review.
Results: 166 patients were eligible for transfer to ACHD services during the study period. 114 successfully transferred to ACHD services, 47 are pending transfer and only 5 (4%) have not attended ACHD clinic as scheduled.
Conclusion: An audit of the ACHD transition program has demonstrated a high rate of successful transfer of young adults to ACHD services with very low rates of loss to follow-up. These results have been achieved with the establishment of a coordinated transition program within paediatric services and the appointment of a dedicated transition CNS working in both paediatric and adult congenital cardiology.

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P1232 - AORTIC STENOSIS IN PREGNANCY OUTCOMES OF A COMBINED CARDIAC & OBSTETRIC ANTENATAL CLINIC

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Introduction: Existing risk scores for assessing risk of pregnancy in women with cardiac disease include left ventricular outflow tract obstruction (LVOTO) as a risk factor for maternal mortality and morbidity. Ante-natal counselling and counselling regarding possible termination of pregnancy is undertaken with these risks in mind. We assessed the maternal and neonatal outcomes of pregnancy in women with significant LVOTO attending our multidisciplinary cardiac antenatal clinic.
Methods: Data was collected for all women with LVOTO with an EDD between April 2015-April 2016. Data collected included diagnosis, complications, medication, mean and peak LVOT gradient at the beginning and end of pregnancy, mode of delivery and birth weight.
Results: We identified 11 pregnant women (age: 20-42yrs) with LVOTO. 7 had moderate LVOTO, and 4 had severe LVOTO. The peak velocity (median +/- IQR) through the aortic valve pre-pregnancy was 3.7 m/s (+/- 0.5 m/s), increasing to a maximum during pregnancy of 4.3 m/sec (+/- 0.8 m/s). Mean valve gradient pre pregnancy was 31 mmHg (+/- 7 mmHg), increasing to a maximum during pregnancy of 40 mmHg (+/-13 mmHg). All babies were born at or around term apart from two babies who were born just before 37 weeks gestation. One patient with severe aortic stenosis and obesity was admitted at 36 weeks with increasing breathlessness. Symptoms settled with rest and introduction of a beta blocker, and she had a successful vaginal delivery at 37 +2/40. 3 women required LSCS for obstetric indications; 8 women had vaginal deliveries. Median birth weight was 3050 g (+/- 1352.5 g).
Conclusion: Outcomes in women with significant LVOTO in our cohort is better than the published risk scores would suggest. Nevertheless, women with LVOTO are potentially at high risk of complications during pregnancy and delivery and should be offered tertiary centre antenatal multi-disciplinary team input. Larger series should be collected to improve accuracy of ante-natal counselling.

P1240 - LONG TERM FOLLOW UP OF REPAIRED TETRALOGY OF FALLOT INTO MIDDLE AGE

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Objective: Survival of patients with repaired tetralogy of Fallot (rToF) into young adulthood is very good. Concerns exist, however, over long term morbidity and mortality as these subjects reach middle age. We aimed to assess survival and the prevalence of complications in rToF patients seen in our Adult Congenital Heart Disease service.
Method: 168 consecutive patients with 'simple rToF', aged over 16 years were included. We documented mortality and analysed the prospectively defined composite endpoints of (a) "Serious adverse events" including death, heart failure hospitalisation and/or documented ventricular arrhythmia and (b) "Adverse events" inclusive of the above and endocarditis, atrial arrhythmia, defibrillator and/or pacemaker implantation.
Results: Mean age at last follow up was 34 ±12 years, 55% were males. There were 10 (6%) deaths and 26 patients (16%) experienced a "serious adverse event". Fifty-one patients (30%) experienced an "adverse event", 29 patients with atrial arrhythmias. One hundred and one (61%) patients had at least one pulmonary valve replacement and 10% required a second pulmonary valve replacement. By age 40 years, 93% of were free of serious adverse events and 83% were free of any adverse event. By age 50 years, only 56% had not had an adverse event. Older age and history of atrial arrhythmia were predictive of serious adverse events.
Conclusion: Survival into mid-adulthood in patients with repaired tetralogy of Fallot is very good, however a substantial number of survivors have clinically significant adverse events by the age of 50 years.

P1241 - LATE DIAGNOSIS OF ADDITIONAL STRUCTURAL ABNORMALITIES IN PATIENTS WITH HISTORIC SURGICAL REPAIR OF ATRIAL SEPTAL DEFECT

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Surgical closure of atrial septal defects (ASD) began in 1953. We report 5 patients who underwent ASD closure years ago, who represent with residual defects. Case 1: Surgical repair ASD 1981 aged 4. 2013, complained of dyspnoea with drop in oxygen saturations to 82% on exertion. Trans-oesophageal echo demonstrated venosus ASD with bi-directional shunting. Patient underwent further surgery to close the residual defect and made a full recovery. Case 2: Surgical repair ASD 1964 aged 14. In 2009 CT demonstrated left pulmonary veins to coronary sinus. Surgically repaired in October 2010. The right heart now normalised in size, but ongoing problems with atrial arrhythmia necessitated drug therapy, two ablations and permanent pacemaker. Case 3: Surgical closure ASD 1971 aged 14. In 2015, CT scan demonstrated drainage of the right sided pulmonary veins to the IVC just above the diaphragm, and a severely dilated right heart. The patient describes poor effort tolerance, and is currently awaiting further surgical intervention. Case 4: Surgical repair ASD 1993 aged 16. Represented 10 years later with paroxysmal atrial arrhythmia, persistently dilated right heart and impaired exercise. Cross sectional imaging demonstrated non-functioning right lung and anomalous drainage of the left upper lobe to innominate vein. The patient underwent further surgery in 2016. Case 5: Surgical repair ASD 1994 aged 18. CT scan in 2014 demonstrated hemianomalous venous drainage of the right lung to the SVC. This patient is currently under consideration for further surgical intervention.
Conclusion: Patients who underwent congenital heart surgery before 2000 did not benefit from modern day imaging techniques and the vast majority are lost to follow up. They should be offered reassessment in specialist services to exclude untreated lesions or late complications. Symptom driven re-referral occurs late in the natural history when irreversible damage may have already occurred.

P1265 - SURGICAL VERSUS TRANSCATHETER DEVICE CLOSURE OF ATRIAL SEPTAL DEFECT (ASD) A COMPARATIVE ANALYSIS OF RIGHT HEART REMODELLING

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Atrial septal defect (ASD) is a common congenital heart abnormality and can remain undetected until adulthood. Closure may be

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attempted even in advanced age, resulting in improvements in symptoms and echocardiographic parameters. Closure is traditionally achieved surgically, with transcatheter device closure gaining popularity in the last few decades.

Aims: The study aims to evaluate the extent and rate of right heart chamber remodeling following transcatheter device closure of ASD in comparison to surgical closure.

Methods: We retrospectively analysed echocardiographic data of 121 patients who underwent either transcatherer device or isolated selective surgical closure of secundum ASD at National Heart Centre, Singapore (NHCS) from 2005 to 2015. Echocardiographic parameters measured pre- and post-intervention included: RA annulus diameter, RV inlet dimension, PASP, TAPSE (tricuspid plane annulus systolic excursion) and S' (systolic velocity of tricuspid annulus).

Results: Patients' age ranged from 18 to 69 years old, with 19 patients of aged 60 and above. There were 57 patients who underwent surgical closure and 64 underwent device closure. Patients who underwent catheter closure were older (46 years vs 37 years, $p < 0.05$). Both groups had similar pre-intervention parameters. A statistically significant decrease in PASP, RV inlet size, RA annulus diameter, S' and TAPSE was observed for both groups at 1 year post-intervention compared to the respective pre-intervention parameters. The biggest change was noted at 6-8 weeks. There was no difference between the two groups in terms of reduction in PASP, RV inlet size and RA annulus. In the device group, the reduction in TAPSE was significantly less up to 1 year with compared to the surgical group.

Characteristic	Total patients n=121
Age (y)	39 (18-69) *Median, (range)*
18-30	38 (31.4%)
31-50	43 (35.5%)
51-70	40 (33.1%)
Sex (Female)	75 (61.9%)
Surgical closure	57 (47.1%)
Transcatheter closure	64 (52.9%)

Figure 1.
Study Population.

Characteristic	Surgical n=57	Device n=64
Age (**P<0.05**)	37 (18-63) (median)	46 (18-69)
Mean systolic BP	122.6	126.8
Mean diastolic BP (mmHg)	70.1	70.3
Mean heart rate (bpm)	76.1	72.0
Baseline PASP (mmHg)	36.0	36.8
RV inlet size (cm)	4.6	4.4
RA annulus diameter (cm)	3.3	3.8
TAPSE (cm)	2.5	2.5
TA TDI S' (cm)	0.14	0.13

Figure 2.
Baseline characteristics: surgical vs device groups.

Conclusions: Device and surgical closure of ASD both result in similar improvement in pulmonary pressures and right heart dimensions. However, device closure is associated with better preservation of TAPSE at 1 year.

Surgical intervention

Characteristic	Baseline (6mo prior)	6-8w		6mo		1y	
PASP	36.0± 12.8	27.4 ± p<0.0001	10.1	25.1 ± p<0.0001	8.7	22.7 ± p<0.0001	8.1
RV inlet size	4.60 ± 0.64	3.89 ± p<0.0001	0.49	3.61 ± p<0.0001	0.43	3.44 ± p<0.0001	0.51
RA annulus diameter	3.27 ± 0.53	2.78 ± p<0.0001	0.40	2.64 ± p<0.0001	0.35	2.56 ± p<0.0001	0.29
TAPSE	2.48 ± 0.44	1.44 ± p<0.0001	0.36	1.82 ± p<0.0001	0.33	1.89 ± p<0.0001	0.26
S*	0.135 ± 0.024	0.089 ± p<0.0001	0.024	0.100 ± p<0.0001	0.018	0.124 ± p=0.565	0.014

Device

Characteristic	Baseline	6-8w		6mo		1y	
PASP	36.4 ± 10.1	29.3± p<0.0001	9.1	25.7 ± p<0.0001	8.3	24.3 ± p<0.0001	7.4
RV inlet size	4.37 ± 0.60	3.84 ± p<0.0001	0.46	3.49 ± p<0.0001	0.39	3.36 ± p<0.0001	0.36
RA annulus diameter	3.25 ± 0.55	2.72 ± p<0.0001	0.38	2.57 ± p<0.0001	0.36	2.54 ± p=0.020	0.39
TAPSE	2.48 ± 0.48	2.25 ± p=0.001	0.37	2.29 ± P=0.008	0.33	2.28± p=0.013	0.35
S*	0.128 ± 0.026	0.127 ± p=0.834	0.025	0.126 ± P=0.722	0.024	0.132 ± p=0.267	0.022

Values are mean ± standard deviation

Figure 3.
Echocardiographic parameters at subsequent follow-ups: comparing surgical vs baseline and device vs baseline at follow-up intervals.

Characteristic	6-8w		6mo		1y	
Change in PASP						
Surgical	-8.6 ±	8.3	-10.9 ±	9.9	-13.3 ±	9.4
Device	-7.2 ± p=0.308	7.2	-10.7 ± p=0.899	9.7	-12.1 ± p=0.5	9.1
Change in RV inlet size						
Surgical	-0.71 ±	0.66	-0.99 ±	0.68	-1.2 ±	0.81
Device	-0.54 ± p=0.145	0.64	-0.88 ± p=0.357	0.63	-1.0 ± p=0.261	0.62
Change in RA annulus diameter						
Surgical	-0.49 ±	0.53	-0.63 ±	0.51	-0.71 ±	0.54
Device	-0.54 ± p=0.650	0.53	-0.69 ± p=0.606	0.59	-0.72 ± p=0.952	0.60
Change in TAPSE						
Surgical	**-1.0 ±**	**0.61**	**-0.67 ±**	**0.56**	**-0.59 ±**	**0.54**
Device	**-0.23 ± p<0.0001**	**0.49**	**-0.19 ± p<0.0001**	**0.56**	**-0.20 ± p<0.0001**	**0.63**
Change in S*						
Surgical	**-0.05 ±**	**0.03**	**-0.03 ±**	**0.03**	-0.01 ±	0.14
Device	**-0.0008± p<0.0001**	**0.03**	**-0.0014± p=0.012**	**0.03**	0.0045 ± p=0.396	0.03

Values are mean ± standard deviation

Figure 4.
Changes in echocardiographic parameters at subsequent follow-ups: Surgical vs Device.

P1279 - QUANTIFICATION OF PULMONARY REGURGITATION BY VECTOR FLOW MAPPING IN CONGENITAL HEART PATIENTS AFTER REPAIR OF RIGHT VENTRICULAR OUTFLOW OBSTRUCTION

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Objective: The new vector flow mapping (VFM) enables direct visualization of flow pattern and estimation of flow volume. We aimed to determine its accuracy in the quantification of pulmonary regurgitation (PR) in congenital heart patients after repair of right ventricular (RV) outflow obstruction.
Methods: This study comprised two parts: i) validation of VFM (Hitachi Aloka, Tokyo, Japan) in the quantification of PR in repaired tetralogy of Fallot (ToF) patients by cardiac magnetic resonance (CMR), and ii) clinical application of VFM to determine PR in patients after biventricular repair of pulmonary atresia (PAIVS) and stenosis (PS) with intact ventricular septum. Pulmonary regurgitation was quantified by calculation of VFM-derived pulmonary diastolic backward flow ratio (PRback), defined as ratio of backward to forward flow volume. In ToF patients, cardiac MRI was performed within 2 months of VFM assessment.
Results: The coefficients of variations for intra- and inter-observer variability in the measurements of PRback were 7.0% and 10.4%, respectively. Fourteen repaired ToF patients aged 31.3 ± 7.3 years were studied. Their PR–back correlated strongly with CMR-derived pulmonary regurgitation fraction ($r = 0.95$, $p < 0.001$) and RV end-diastolic volume ($r = 0.84$, $p < 0.001$). In the second part, 14 PAIVS patients aged 25.6 ± 6.0 years, 14 PS patients aged 24.2 ± 7.0 years, and 14 healthy controls were studied. The VFM-derived PR–back was found to increase across groups of subjects with absent (4.6 ± 3.3%), mild (11.1 ± 7.1%), moderate (29.6 ± 7.8%) and severe (50.1 ± 8.2%) PR as defined semi-quantitatively by colour flow mapping. Furthermore, VFM-derived PR–back correlated strongly with the ratio of PR colour jet/width of RV outflow ($r = 0.92$, $p < 0.001$).
Conclusion: Vector flow mapping is a reproducible technique for accurate quantification of PR in congenital heart patients after repair of right ventricular outflow obstruction.

P1288 - COMPOUND HETEROZYGOUS LOSS OF FUNCTION MUTATIONS IN KIF20A ARE ASSOCIATED WITH A NOVEL LETHAL CONGENITAL CARDIOMYOPATHY IN TWO SIBLINGS

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Aims: Congenital or neonatal cardiomyopathies are commonly associated with a poor prognosis and have multiple etiologies. In two siblings, a male and female, we identified an undescribed type of lethal congenital restrictive cardiomyopathy affecting the right ventricle. We hypothesized a novel autosomal recessive condition. To identify the cause, we performed genetic, in vitro and in vivo studies.
Methods and Results: Genome-wide SNP typing and parametric linkage analysis was done in a recessive model to identify candidate regions. Exome sequencing analysis was done in unaffected and affected siblings. In the linkage regions, we selected candidate genes that harbor two rare variants with predicted functional effects in the patients and for which the unaffected sibling is either heterozygous or homozygous reference. We identified two compound heterozygous variants in KIF20A; a maternal missense variant (c.544 C > T: p.R182W) and a paternal frameshift mutation (c.1905delT: p.S635Tfs*15). Functional studies confirmed that the R182W mutation creates an ATPase defective form of KIF20A which is not able to support efficient transport of Aurora B as part of the chromosomal passenger complex. Due to this Aurora B remains trapped on chromatin in dividing cells and fails to translocate to the spindle midzone during cytokinesis. Translational blocking of KIF20A in a zebrafish model resulted in a cardiomyopathy phenotype.
Conclusion: We identified a novel autosomal recessive congenital restrictive cardiomyopathy, caused by a near complete loss-of-function of KIF20A. This finding further illustrates the relationship of cytokinesis and congenital cardiomyopathy.

P1300 - NON OSMOTIC ARGININE VASOPRESSIN SECRETION IN ADULT PATIENTS WITH CONGENITAL HEART DISEASE

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Background: Recently, the cardio-renal connection is receiving a lot of attention in management of heart failure. Arginine vasopressin (AVP) system is one of the most important systems to connect heart and kidney. However, the knowledge about the relationship between the AVP system and the hemodynamics is limited. There are two mechanisms in AVP secretion, osmotic and non-osmotic control, and the change of hemodynamics could influence on the AVP secretion via non-osmotic control. The purpose of this study is to clarify the relationship between the non-osmotic AVP secretion and the hemodynamics in adult patients with congenital heart disease (CHD).
Methods: This study enrolled 74 adult CHD patients. Peripheral blood was drawn in fasting for analysis of plasma osmolality and AVP concentration. Using the osmolality, the estimated AVP concentration was calculated. A condition with higher measured AVP level than estimated AVP concentration was defined as the elevation of the non-osmotic AVP secretion. The various parameters were compared between the patients with and without the elevated non-osmotic AVP secretion.
Results: The patients' age was 35.2 ± 14.8 years. NYHA class was I:46, II:21, III:7, IV:0. 17 patients suffered from cyanosis. The level of brain natriuretic peptide was 81.4 ± 174.0 pg/ml. The plasma AVP level was 2.3 ± 2.1 pg/ml and osmolality was 291.9 ± 3.6 mOsm/l. The non-osmotic AVP secretion elevated in 10 patients and the AVP level (6.4 ±3.1 pg/ml) of them was higher than that in the patients without the elevation (1.7 ± 0.9 pg/ml). Diuretics use was significantly frequent in the elevated non-osmotic AVP secretion group (60% vs. 16%; Odds Ratio 8.10 (1.93-33.98), $p = 0.0053$).
Conclusions: In adult patients with CHD, diuretics administration is one of the causes of the elevation of the non-osmotic AVP secretion. Volume depletion by diuretics may induce the non-osmotic AVP secretion.

P1304 - CAN THE USE OF A FITBIT INCREASE ACTIVITY IN ADULT CONGENITAL PATIENTS WITH PULMONARY HYPERTENSION

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Introduction: Little is known on the activity levels of adults with pulmonary hypertension in association with congenital heart disease. We sought to quantify activity levels in this group with FitBit

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exercise trackers and used the data to motivate individuals to increase their activity levels.
Method: 18 patients were given a FitBit and joined a closed online group. Activity data (daily number of steps) were collected over 3 months. After collecting baseline data for 2 weeks, feedback was given to individuals and they were each asked to try to increase their activity by 10%. Regular contact and encouragement were provided through the online group, email and phone. Formal feedback was collected at 3 months.
Results: The patient group ranged from 19 years to 69 years, 11 were female & 7 male, 6 had Trisomy 21. All patients completed the programme for the full 3 months. Mean baseline daily activity was 4566 steps (range 1833 to 8792). Daily step counts increased by 13% at 4 weeks, 13.9% at 8 weeks and 10.9% at 12 weeks compared to baseline. 10 of 18 patients completed the feedback questionnaire: 20% stated that they felt much better, 60% a little better and 20% about the same. Comments from patients included: "made me feel better", "increased confidence", "has given me something positive to focus on."
Conclusion: Activity levels improved in our patients with the use of an activity tracker supported by encouragement from health professionals. The effect was sustained at 3 months. There also appeared to be a positive effect on patient-reported well-being. The programme continues and we will continue to evaluate effectiveness over the longer term.

P1329 - VENTRICULAR ASSISST DEVICE IN ADULT CONGENITAL HEART DISEASE A SYSTEMATIC REVIEW AND META ANALYSIS

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Introduction: Advanced heart failure is an increasing problem in adult congenital heart disease (ACHD) patients. Data regarding survival after ventricular assist device (VAD) placement in ACHD are scarce. We reviewed current literature to assess survival in ACHD patients with VADs.
Methods: A systematic review was performed utilizing Pubmed, Ovid, and Cochrane for all reported ACHD patients (age >21) who received VADs. Data on baseline demographics, duration of VAD support, BTT (Bridge to Transplant) vs DT (Destination Therapy), ACHD lesion, death, and transplantation were collected. Kaplan-Meier estimates were used to assess survival.
Results: Literature review yielded 43 ACHD patients who underwent VAD implantation (mean and median age 41 yrs, range 23-75 yrs.). Duration of VAD support: mean 329 days (median 244, range 7-1477). Gender was reported in 39 patients (33 males). VAD was placed as DT in 11 pts, of whom 2 died. BTT was the indication in 32 pts: 16 were transplanted, 7 pts died, and 9 were still awaiting transplant on VAD support. DTGA (n = 19) and ccTGA (n = 18) were the most commonly reported ACHD lesions. Single ventricle S/P Fontan was reported in 5 pts: 2 were transplanted, 2 were DT and one died awaiting transplant at 262 days. Continuous-flow VADs were more commonly used (n = 35) vs 7 pulsatile-flow, 4 VAD types were not reported. Most VADs were implanted to the morphologic RV (n = 38), 8 to the LV including 3 BIVADS. The 30 day and 1 year survivals post-VAD placement in ACHD patients were 88.5% and 80.5% respectively with a survival estimate of 70.5% at 2 years. This is similar to INTERMACS reported 1 year survival of 80% and 2 year survival of 70% with CF devices in all patients.
Conclusions: In carefully selected ACHD patients, VADs can be successfully used for mechanical circulatory support.

P1347 - A SUCCESFUL SURGICAL REPAIR OF TETRALOGY OF FALLOT IN ADULT WITH DEXTROCARDIA AND SITUS INVERSUS TOTALIS

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Background: Tetralogy of Fallot, a combination of four related heart defects, is the most common cyanotic heart disease in Indonesia. Dextrocardia is a condition in which the major axis of the heart points to the right and associated with intracardiac lesion. However, Tetralogy of Fallot in patient with dextrocardia and situs inversus is rare and present technical challenges in the surgical repair. The surgical repair of dextrocardia with congenital lesion is easier to performed from the left side.
Purpose: To report a successful surgical treatment in adult with Tetralogy of Fallot, situs inversus totalis and dextrocardia that previously had imprecise preoperative diagnosis of pulmonary stenosis with vegetation and infective endocarditis.
Method: A 22 years old girl was referred to our hospital with shortness of breath and history of recurrent syncope. The diagnosis of situs inversus totalis and dextrocardia with severe supravalvular pulmonal stenosis, moderate pulmonal regurgitation were established, based on chest radiograph, transthoracal echocardiography and cardiac CT scan. The patient was also diagnosed as infective endocarditis and has got antibiotic for a month. Patient was originally planned to underwent pulmonary valve replacement. During surgery, transesophageal echocardiography evaluation confirmed the diagnosis of Tetralogy of Fallot.
Discussion: During surgery, we found the heart located in the right hemithorax with its base to apex axis directed to the right, superior and inferior vena cava at the left side, and right aortic arch. After opening right atrium, there were patent foramen ovale, infundibular thickening, and subaortic ventricular septal defect at diameter 1 cm without vegetation. Infundibular thickening was resected, ventricular septal defect was closed with dacron patch and transanullar patch was done with pericardium graft.
Keywords: Tetralogy of Fallot, situs inversus totalis, dextrocardia, pulmonal stenosis, ventricle septal defect (VSD), patent foramen ovale (PFO).

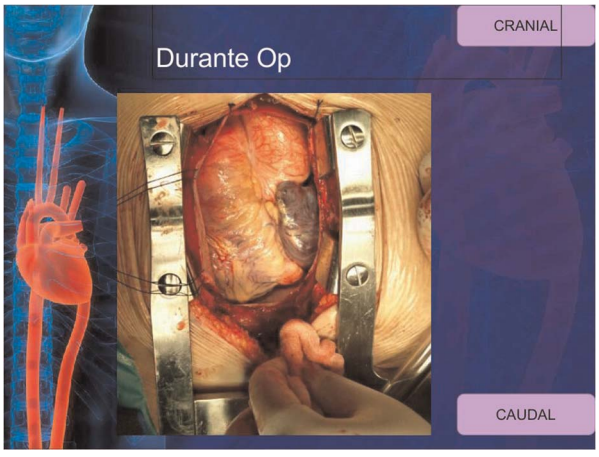

Figure 1.

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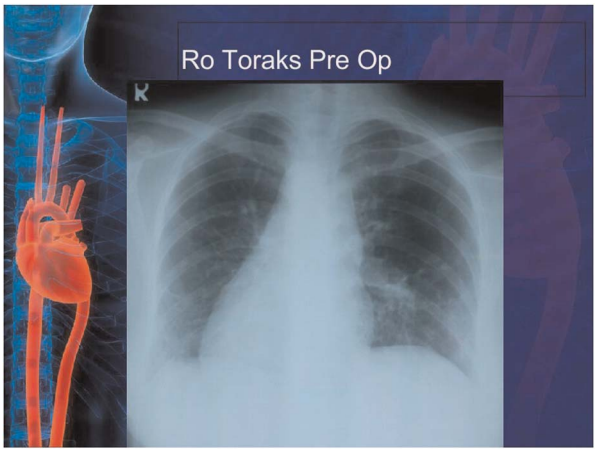

Figure 2.

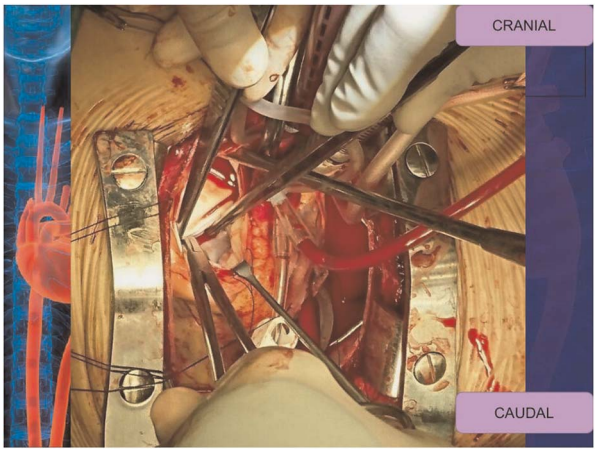

Figure 3.

P1366 - INCIDENCE AND OUTCOME OF INFECTIVE ENDOCARDITIS FOLLOWING PERCUTANEOUS VERSUS SURGICAL PULMONARY VALVE REPLACEMENT

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Background: Transcatheter pulmonary valve replacement (TPVR) may be associated with a higher risk of infective endocarditis (IE) than surgical pulmonary valve replacement (SPVR), however, there is a paucity of data to support this assertion.
Materials and Methods: Patients who underwent TPVR or SPVR at UCLA between October 2010 and September 2016 were included. Baseline and subsequent clinical data were retrospectively collected. IE was diagnosed based on the modified Duke criteria.
Results: A total of 342 patients underwent pulmonary valve replacement at UCLA including 134 SPVR and 208 TPVR. Patients in the TPVR group were more likely to have had a history of endocarditis (5.3% vs 0.7%, P = 0.03) and more likely to have a right ventricle to pulmonary artery (RV to PA) conduit (37% vs 17%, P = 0.0001). Two SPVR (1.5%) and eight TPVR cases (3.8%, P = 0.21) developed IE following the procedure with a 5-year freedom from endocarditis of 94.0% in the SPVR versus 83.8% in the TPVR group (P = 0.08). In patients who underwent TPVR and developed endocarditis, the mean gradient across the RVOT prior to intervention was significantly higher when compared to the patients who underwent TPVR and did not develop endocarditis (28.1 ± 4.5 mmHg vs 17.4 ± 0.6, P = 0.02). In addition, patients who developed endocarditis were more likely to have an RV to PA conduit when compared to patients who did not develop endocarditis.
Conclusions: There is a trend that suggests that patients undergoing TPVR may be at a higher risk of IE than patients undergoing SPVR. TPVR patients were far more likely to have had a prior history of IE and a conduit. The patients at highest risk are those with stenotic RV to PA conduits who are treated with TPVR.

P1376 - HEART FAILURE ADMISSIONS AND POOR SUBSEQUENT OUTCOMES IN ADULTS WITH CONGENITAL HEART DISEASE

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Background: Heart failure is a leading cause of morbidity and mortality amongst adults with congenital heart disease (CHD). Outcomes following hospital admission with heart failure in adults with CHD, however, are largely unknown.
Aim: To determine the prevalence, complications and prognosis of clinical heart failure in a cohort of patients with adult congenital heart disease (ACHD).
Methods: Data on hospital admissions for heart failure (HF) were collected from our tertiary referral centre between 2000 and 2014. We analysed CHD diagnosis, survival status and cause of death. Vital status was known for all subjects from a National Death Registry. We also gathered information on subject demographics, co-morbidities, medical treatment and ventricular function (based on echocardiography).
Results: Data from 3862 consecutively registered and qualifying adult CHD patients were collated. 179 (4.6%) patients had been admitted to hospital with a clinical diagnosis of heart failure, with a mean age of 57 years. HF was more likely in those with more complex defects or with bicuspid aortic valve disease. HF was also more common in those with concomitant atrial arrhythmias and/or severe left ventricular dysfunction. 5-year mortality rates were 45% for patients in the HF group and 8% for the ACHD patients with no HF admissions ($p < 0.001$). More than 70% of the HF patients died within 10 years of their first admission, with 25% dying in the first year. Over 90% of these deaths were attributable to cardiovascular causes.
Conclusion: Admission to hospital with heart failure is a strong, negative prognostic event in patients with ACHD. It is associated with high rates of mortality, especially in the first year following hospital admission. As such, there is a need to identify modifiable risk factors to prevent first (and subsequent) heart failure admission(s).

P1378 - USEFULLNESS OF EUROSCORE II TO PREDICT COMPLICATION OF CARDIAC RE OPERATION AMONG ADULT TETRALOGY OF FALLOT PATIENTS; A SINGLE CENTER EXPERIENCE

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Background: Cardiac re-operation is sometimes needed among adult tetralogy of Fallot (TOF) long-term after repair. Risk factor of re-operation among TOF patients is not clarified. EuroSCORE II, which can predict mortality of adult cardiac surgery, is calculated based on risk factors (NYHA, LVEF, weight of procedure et cetera) and their coefficient.
Objective: The aim of this study is to investigate utility of EuroSCORE II for predicting complications of re-operation among TOF patients.
Methods: A retrospective review identified 26 adult TOF who underwent re-operation at Chiba Cardiovascular Center from January 2008 to November 2016. Patients were classified into two groups; those with perioperative complications (Group A) and those without (Group B). Perioperative complications were defined as follows; A death within 30 days, cardiac failure requiring mechanical circulatory support, renal failure requiring dialysis, cerebral infarction or bleeding, myocardial infarction, intubation/NIPPV over 48 hours, length of ICU stay over 6 days, re-operation. We compared two groups about following indices; predicted mortality calculated by EuroSCORE II and items which EuroSCORE II doesn't include (RVEDVI, RVEF, co-existence of PA or MAPCAs).
Results: The breakdown of complications was 2 re-operation, 5 intubation/NIPPV over 48 hours, 4 long ICU stay (duplicates exist). Predicted mortality was significantly high in Group A than Group B($p = 0.026$)(Group A; median 11.7%: range 4.3-46.5%), Group B; median 5.2%: range 2.7-9.1%). There were no significant differences between two groups about other indices; index: median(range) or proportion of Group A and B, p value. RVEDVI(ml/m2): 147(range 98-320), 144(range 71-323), $p = 0.53$, RVEF(%): 34(range 24-47), 45(range 21-59), $p = 0.11$, Co-existence of PA: 25%, 10%, $p = 1$, Co-existence of MAPCAs: 0%, 14%, $p = 1$.
Conclusion: EuroSCORE II can be applied to predict perioperative complications of re-operation among adult TOF patients, and further study is required to develop tools that predict complications of re-operation in them.

P1382 - A CLUSTER RANDOMIZED TRIAL OF A TRANSITION INTERVENTION FOR ADOLESCENTS WITH CONGENITAL HEART DISEASE

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Background: The population of adults with congenital heart disease (CHD) is growing exponentially. However, there is little evidence regarding the preparation of adolescents to assume responsibility for their health and self-management within the adult health care system.
Methods: We conducted a randomized clinical trial of a nurse-led transition intervention for 16-17 year olds with moderate or complex CHD, versus usual care. The intervention group received two 1-hour individualized sessions with a cardiology nurse, focusing on CHD education and self-management skills. The primary endpoint was excess time to adult CHD care, defined as the interval between the final pediatric and first adult cardiology appointments, minus the recommended time interval between these appointments. Secondary endpoints were MyHeart score (CHD knowledge) and Transition Readiness Assessment Questionnaire (TRAQ) score.
Results: 121 participants were randomized to intervention (57) or usual care (64). Freedom from excess time to adult CHD care was similar in both groups (log rank $p = 0.09$). At the recommended time of first adult cardiology appointment (excess time = 0), intervention participants were 1.78 times more likely to have their adult cardiology appointment within one month relative to those in usual care (95% CI 1.11-2.86, Cox regression $p = 0.018$). This hazard increased with time; at an excess time of 6 months, intervention participants were 2.97 times more likely to have an appointment within one month (95% CI 1.07–8.26). MyHeart scores were higher in the intervention group compared to usual care at 1, 6, 12 and 18 months ($p < 0.001$, mixed models), as were TRAQ self-management scores ($p = 0.032$, mixed models).
Conclusions: A nurse-led intervention program reduced the likelihood of a delay in obtaining adult CHD care after graduation from pediatric cardiology and resulted in greater CHD knowledge and self-management skills up to 18 months post intervention. A structured intervention program should be provided to all adolescents with CHD.

P1430 - BICUSPID AORTIC VALVE PHENOTYPE AFFECTED VALVULAR FUNCTION AND AORTIC ROOT GEOMETRY IN CHILDREN AND YOUNG ADULTS

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Objective: In adult patients, the relationship of phenotype of bicuspid aortic valve (BAV) between severe aortic stenosis (AS) or regurgitation (AR) has been reported. However, the relationship in children was not documented well. Therefore, we evaluated the aortic valve dysfunction according to BAV phenotype in pediatric patients.
Methods: Between Jan. 2005 and Dec. 2014, 89 BAV patients were reviewed retrospectively. According to BAV morphology, patients were divided into two groups; BAV-R/N (right coronary cusp (RCC) and non-coronary cusp fusion) and BAV-R/L (RCC and left coronary cusp fusion). Demographics, size of ascending aorta (AA), aortic valve function, and event of surgery or intervention were compared between two groups.
Results: The median age of 89 patients was 3.5 (range, 0-17.8) years. The number of BAV-R/N was 52 (58.4%), and there was no age and sex difference between groups. AS was more frequently associated with BAV-R/N (55.8% vs 29.7%, $p = 0.013$). Aortic root dilatation was more prevalent in BAP-R/L group (7.7% vs 29.7%, $p = 0.017$). Sinus of Valsalva Z-score was consistently larger in BAV-R/N group. During follow up, two (2.2%) patients underwent aortic valvuloplasty for AR, and seven (7.6%) patients underwent intervention (n = 5) or surgery for AS. Aggravated aortic root and AA dilatation was occurred in five and six patients, respectively.

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Conclusion: BAV is frequently associated with AS, AR and AA dilatation. AR could occur in young adults. Therefore, complete follow-up is warranted. Recognition of BAV morphology is helpful for predicting of the type of aortic valve dysfunction and the location of AA dilatation.

P1431 - VENTRICULAR MORPHOLOGY IN ADULTS BORN WITH A VENTRICULAR SEPTAL DEFECT

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Background: Ventricular septal defects (VSD) – whether they are large and surgically closed or small and untreated – are considered to have great outcomes in adulthood. However, we have previously demonstrated lower functional capacity in both patient groups compared with healthy, matched controls. The mechanisms behind these findings remain unclear and therefore, we performed cardiac magnetic resonance imaging (MRI) on adult patients in order to evaluate biventricular morphology.
Methods: Adults with surgically closed or small, untreated VSDs and healthy controls underwent cine MRI for the evaluation of the biventricular volumes, and phase contrast scans for the measurement of cardiac index. MRI measurements were analysed post hoc in a blinded fashion by one main investigator.
Results: Twenty operated patients (22.4 ± 2years) and 20 matched controls (23.1 ± 2years) were included, along with 32 small, open VSDs (26.3 ± 6years) and 28 matched controls (26.8 ± 5years). Operated VSDs were found with larger right ventricular end-diastolic volume index (RVEDVi) (102.7 ± 20 ml/m2) compared with their controls (90.4 ± 15), $p = 0.03$. Heart rate and right ventricular cardiac index (RVCi) were comparable between operated VSDs and controls. Open VSDs also revealed larger RVEDVi (105 ± 17 ml/m2) compared with their matched controls (88.2 ± 12 ml/m2), $p < 0.01$. Furthermore, RVCi was higher in patients with open VSDs (4.4 ± 1.2 l/min/m2) compared with controls (3.6. ± 1 l/min/m2), $p < 0.01$, but heart rates were similar. Left ventricular measurements displayed no differences between patient groups and their matched control groups.
Conclusion: An altered RV morphology was demonstrated in adults with small, untreated VSDs and in adults 20 years following surgical VSD-repair. These findings may explain some of the mechanisms behind the exercise limitations previously found in adulthood in this patient group.

P1448 - IMPAIRED CARDIAC OUTPUT AND INCREASED RETROGRADE PULMONARY FLOW DURING EXERCISE IN ADULTS OPERATED FOR VENTRICULAR SEPTAL DEFECT IN CHILDHOOD

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Background: The long-term outcome after surgical closure of ventricular septal defect (VSD) in childhood is not as benign as previously believed. Recent studies have shown markedly reduced functional capacity and disruption of the right ventricular force-frequency relationship during exercise in adult survivors of VSD. In order to further describe the long-term cardiac function, we performed a non-invasive assessment of cardiac index during exercise in adults operated for ventricular septal defect in early childhood.
Materials and Methods: Twenty patients (surgical age 2.1 ±1.4 years, age at examination 22.1 ±2.2 years) and twenty healthy, matched controls (23.4 ±2.1 years at examination) underwent continuous supine bicycle ergometry during magnetic resonance imaging. Blood flow was recorded in the ascending aorta and the pulmonary trunk at increasing exercise levels. Cardiac index, retrograde flow and vessel diameter were determined by blinded analyses.
Results: Mean cardiac index was comparable at rest with 2.9 ±0.7 l/min/m2 in the patient group and 3.0 ±0.6 l/min/m2 in the control group. During exercise, the patients had a lower increase in cardiac index reaching 7.3 ±1.3 l/min/m2 at sub-maximal exercise compared with 8.2 ±1.2 l/min/m2 in controls, $p < 0.05$. In the ascending aorta, patients had significantly higher retrograde flow than the controls at rest and throughout exercise. In the pulmonary artery, retrograde flow was minimal at rest in both groups, but increased significantly in the patient group during exercise compared with the control group. The patients had significantly larger aortic diameter of 32 ±5 mm compared with 28 ±3 mm in controls, $p < 0.01$. There was no difference in pulmonary trunk diameter.
Conclusions: Young adults with a surgically closed VSD have reduced cardiac index during exercise when compared to healthy young adults. The impaired cardiac index appears to be related to increasing retrograde flow in the pulmonary artery with progressive exertion.

P1450 - ECHOCARDIOGRAPHY MYOCARDIAL FUNCTION CORRELATE WITH CHANGE IN CARDIAC VARIANT FABRY DISEASE

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Background: Fabry disease (FD) is an X-chromosomal recessive deficiency of the lysosomal hydrolase alpha-galactosidase A (alpha-Gal), which is related to the IVS4 mutation and is used to treated with recombinant enzyme replacement therapy (ERT). Although left ventricular (LV) hypertrophy is the hallmark of cardiac involvement, its underlying pathologic represents of the total cardiac mass is not fully understood.
Methods: We enrolled 10 patients, 7 males and 3 females, median age was 59 who hobored cardiac variant Fabry disease in Taiwan. Firstly, we detected phosphatase and tensin homolog gene (PTEN) rs3830675, VS4 polymorphism in DNA samples extracted from whole blood by using DNA mass spectrometry. To use cardiac magnetic resonance imaging (MRI) and echocardiography for criticizing cardiac fibrosis and LV diastolic dysfunction, respectively. Histologically, we performed the estimation on inter-cell fibrosis and cell replacement by using haematoxylin and eosin (H & E) and Masson's trichrome and touluidine blue. On the other hand, the periodic acid-Schiff (PAS), lyosome-associated membrane protein-2 (LAMP2), CD31, CD34, GLA and globo-triaosylceramide (Gb3) of histochemistry in endomyocardial biopsy of these patients were also performed.
Results: A total of 10 cardiac variant Fabry disease of patients had intronic mutation IVS4 + 919G- > A ,and found LV diastolic dysfunction. Importantly, we found PAS (≦ 20%), cardiac muscle cell nucleus size (>6 μm), Vacuolization (>50%) were positively correlated with moderate to severe LV diastolic dysfunction, and

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two cellular cluster differentiation markers CD 31 and 34 expressions were not associated with LV diastolic dysfunction. However, we used tranditional Electrocardiogram (ECG) to confirmed these 10 cardiac variant Fabry of patients was found one patient with normal ECG had short run of PSVT and triplet VPCs via 24-hour holter monitoring.

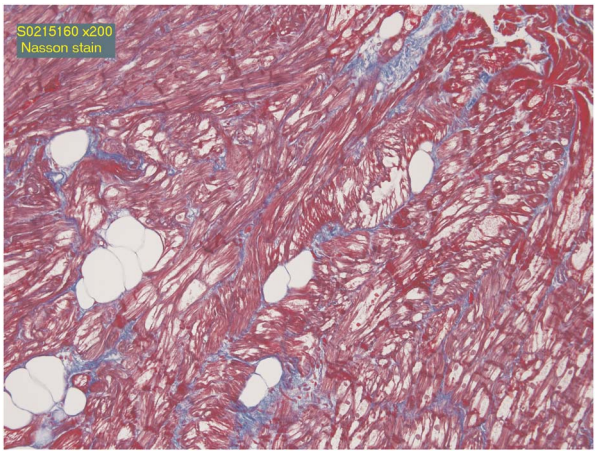

Figure 1.

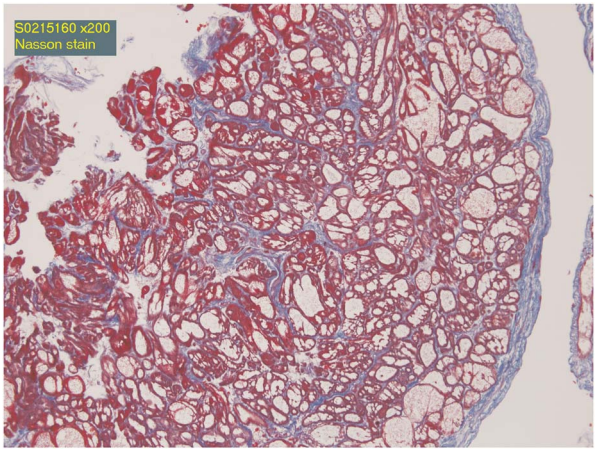

Figure 2.

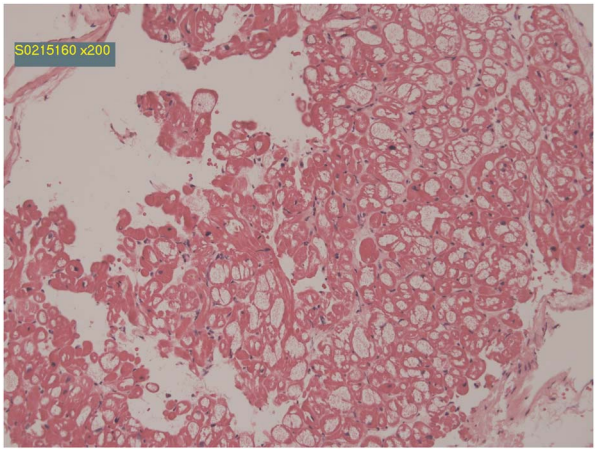

Figure 3.

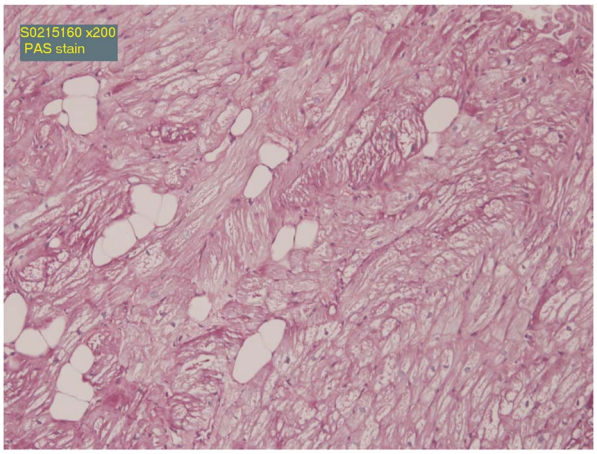

Figure 4.

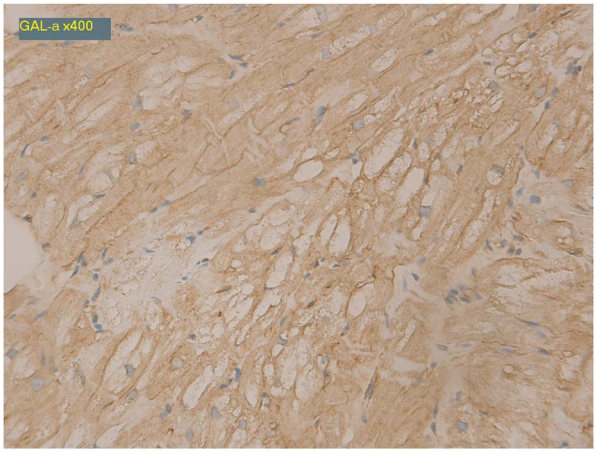

Figure 5.

P1451 - OUTCOME OF BIOPROSTHETIC VALVES IN PULMONARY POSITION IMPLANTED FOR RECONSTRUCTION OF THE RIGHT VENTRICULAR OUTFLOW TRACT IN ADULT CONGENITAL HEART DISEASE

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Objectives: Pulmonary valve replacement (PVR) is the most frequent surgical procedure in adult patients with congenital heart diseases (ACHD). However, none of used valve substitutes is ideal. The aim of the study was to determine the impact of PVR on clinical outcome and right ventricular function.
Methods: We retrospectively reviewed the outcome and performance of bioprosthetic valves implanted in the pulmonary position. Since 2005 to April 2015, 133 consecutive patients with a median age of 35 (range 18.4 to 70.4) years underwent valve implantation: Carpentier-Edwards Perimount MagnaTM (N = 106), St. Jude TrifectaTM (N = 10), St.Jude Medical EpicTM (N = 11), St. Jude TorontoTM (N = 4) and other (N = 2). Median follow-up was 4.3 (0.1-11.0) years. Patients

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diagnoses included tetralogy of Fallot (n = 84), pulmonary stenosis (n = 33), complex diagnoses (n = 14) or other (n = 2). One hundred twenty nine patients had 182 previous surgical procedures. Eighty-two patients (61%) had concomitant surgical procedures.
Results: There were no perioperative deaths, and 1 (0.8%) late death. Probability of freedom from reoperation was 100%, 99%, and 86.1% at 1, 4, and 8 years, respectively. Probability of freedom from valve dysfunction (pulmonary insufficiency more than moderate and or stenosis >40 mmHg) was 98.3%, 90.2%, and 56.9%, respectively. There was a significant improvement ($p < 0.001$) in NYHA functional class. Echocardiography confirmed lasting reduction in pulmonary ($p < 0.001$) and tricuspid valve regurgitation ($p < 0.001$), decrease in the size of tricuspid valve annulus ($p < 0.001$) and improvement of right ventricular function ($p < 0.001$).
Conclusions: PVR using bioprosthetic valves has a low mortality and carries lasting improvement in functional status and right ventricular function in ACHD patients. Freedom from re-operation and valve dysfunction is acceptable. Further studies are needed to compare long-term performance of different valves types in the pulmonary position. (Supported by MH CZ - DRO, University Hospital Motol, Prague, Czech Republic 00064203)

P1463 - CLINICAL AND HEMODYNAMIC FEATURES OF EISENMENGER SYNDROME A TERTIARY REFERRAL CENTER EXPERIENCE

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Background/Hypothesis: Eisenmenger syndrome (ES) is the most severe form of pulmonary arterial hypertension and arises on the basis of congenital heart disease with a systemic-to-pulmonary shunt. We aim to investigate characteristics and outcomes of ES patients at a tertiary center.
Materials and Methods: The data of 64 patients (mean age = 39 years, 32 male and 32 female, mean follow-up time = 20.6 years) with ES were reviewed retrospectively. Demographic characteristics, symptoms, laboratory and hemodynamic parameters, and medications were analyzed.
Results: The ES was diagnosed at 19.5 ± 13.1 years and underlying congenital heart disease was confirmed at 13.6 ± 13.9 years. The most frequent underlying heart diseases were isolated ventricular septal defect and patent ductus arteriosus (n = 17, 22%; respectively). 48 patients (77%) of ES were undergone cardiac catheterization (mean pulmonary arterial pressure = 73.5 ± 17.7 mmHg, arterial oxygen saturation = 87.4 ± 6.1%). Dyspnea (n = 45, 70%) and polycythemia (n = 26, 40%) were common, and hemoptysis (n = 14, 22%) and brain abscess (n = 6, 9.3%) were frequent complications. Functional capacity of NYHA Class III-IV and II was 22% and 62%, respectively. Pulmonary arterial hypertension specific drugs were used in 22% patients at 27.3 ± 5.5 years. Among 9 patients with pregnancy, 5 women underwent delivery without complication. Mortality was 10 patients (15.6%) and aggravating factors were as follows: heart failure, infection, sudden death, and delivery.
Conclusions: Although there are various complications and poor functional capacity, ES patients survive into their third or fourth decades. Therefore more active treatment and clinical follow up might be helpful for their quality of life.

P1473 - THE OUTCOME OF TRANSCATHETER COIL OCCLUSION OF FENESTRATION AFTER TOTAL CAVOPULMONARY CONNECTION

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Fenestration of conduit of total cavopulmonary connection (TCPC) have patients of high risk. Risk factors included pulmonary artery (PA) pressure of 15 mm Hg or more, end-diastolic pressure of 12 mm Hg or more, valve regurgitation, significant cardiac arrhythmias, stenosis or hypoplasia branches PA, pulmonary vascular resistance of 2 Woods' units or more, ventricular outflow obstruction. We studied 10 patients 14.4 ± 7.6 years of age after TCPC operation.
Objectives: To determine the effects of closure of fenestration of conduit of TCPC on hemodynamics and clinical outcome of patients.
Methods: From 2003 to 2016 in UCCC 132 patients were operated TCPC. All patients had high preoperative risk before TCPC. Patients presented with moderate cyanosis, clubbing, effort intolerance (NYHA class II). Surveys revealed a hemoglobin level of 19.0 ± 10 gm•dL-1, hematocrit of 55 ± 5%, and peripheral saturation of 80 ± 5%. Fenestrations were occluded with a balloon to determine the effects on hemodynamic: right atrial pressure was acceptable (<20 mm Hg), aortic pressure remained stable with test occlusion Contraindication to the closure of fenestration: increase pressure in the Fontan anastomosis 3-5 mm Hg, an increase in difference arterio-venous blood by 10% higher, reducing the systemic systolic blood pressure 10% and above.
Results: Average postoperative follow-up was 18.8 ± 5.8 mon. During the follow-up visit the clinical condition of the patients was examined, tests with exercises stress were conducted to assess functional status, the level of blood oxygen saturation and hemoglobin, evaluated data of ECG, Echo, chest X-Ray. All patients reported improved exercise tolerance (NYHA I). Hemoglobin level of 16.5 ±5 gm•dL-1, hematocrit of 45 ±5%, and peripheral saturation of 95 ±2%.
Conclusions: Percutaneous occlusion of fenestration – it effective and safe method of closing the shunt in patients after TCPC. After the procedure, patients noted improvement:the absence of complaints, increased saturation and reducing the level of hemoglobin.

P1483 - HEART RATE RESPONSE TO EXERCISE IS IMPAIRED IN ADOLESCENTS AND YOUNG ADULTS WITH CONGENITAL HEART DISEASE COMPARED TO HEALTHY CONTROLS

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Background/Hypothesis: Adolescents and young adults with congenital heart disease (CHD) frequently have impaired exercise capacity. Purpose of this investigation was to assess heart rate response to exercise and its impact on exercise capacity in adolescents and young adults with CHD compared to healthy controls.
Materials and Methods: 192 adolescents and young adults (128 CHD with varying diagnoses (mean age [+/-SD] 14.52 +/-4.75 years) and 64 age-matched controls (14.23 ± 6.55 years) having performed cardiopulmonary exercise testing were included in this

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retrospective analysis. Patients treated with beta-blockers or with pacemakers were excluded. Peak oxygen consumption (peak VO2), resting heart rate, and the increase in heart rate from resting level to peak exercise (heart rate reserve) were measured. Chronotropic incompetence (CI) was defined as the inability to achieve 80% of age predicted maximal heart rate reserve ([peak heart rate-resting heart rate]/[220 − age − resting heart rate]).
Results: Peak VO2/kg was significantly lower in CHD compared to controls (34.49 ± 7.96 vs 40.87 ± 7.55 ml/kg/min; p <0.0001). Resting heart rate was similar in CHD compared to controls (93.95 ±12.59 vs 97.65 ± 15.02; p = 0.07). Heart rate reserve during exercise was 80.26 ± 13.89 in CHD compared to 86.29 ± 18.71 in controls (p = 0.02). The prevalence of CI was 67% in patients with CHD compared to 48% in controls (p = 0.001), and was most impaired in patients with Fontan circulation (94%; p = 0.0005). Patients with CI had a lower peak VO2/kg (35.13 ± 8.91 vs 38.95 ± 6.87 ml/kg/min, p = 0.002), and shorter exercise duration (11:43 ± 02:02 vs 13:00 ± 01:20 min; P < 0.0001) than those without CI.
Conclusions: Adolescents and young adults with CHD show an impairment of heart rate response to exercise compared to healthy controls. According to our results, chronotropic incompetence is common, especially in patients with Fontan circulation, and correlates with exercise limitation.

P1486 - DETERMINANTS OF CARDIAC OUTPUT IN ADULT PATIENTS WITH FONTAN CIRCULATION

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Background: Low cardiac output (CO) is one of major hemodynamic characteristics of pediatric patients after Fontan operation and may be associated with poor prognosis. However, preserved CO is not always a better hemodynamic phenotype in adult Fontan patients and the clinical determinants have not been clearly demonstrated.
Methods: We prospectively estimated cardiac index (CIdye) and standardized circulating blood volume (BV: ml/kg) during catheterization in consecutive 57 Fontan patients and 19 patients with biventricular physiology (BVP), mostly tetralogy of Fallot based on a pulse dye dilution method with an injection of indocyanine green (ICG) into the pulmonary artery. Results were compared with the hemodynamics obtained by conventional Fick method.
Results: BV was grater in patients with Fontan than those with BVP (72 ± 15 vs. 94 ± 32, p = 0.005), while no difference in CI (p = 0.89). In Fontan patients, CIdye was correlated with CI obtained by Fick method (r = 0.604, p < 0.0001). According to the univariate analysis, low systemic vascular resistance (Rs), arterial oxygen saturation (SaO2) and plasma levels of albumin, high hepatic venous wedge pressure (HVWP) and BV were associated with increased CIdye (p < 0.05-0.0001). Of those, increased BV (p < 0.0001) and low Rs (p < 0.05) were independently associated with the increased CIdye. On the other hand, CIdye showed no relationship with ejection fraction and end-diastolic volume index of the systemic ventricle.
Conclusion: CO is mainly determined by BV and Rs in adult Fontan patients and the hepatic dysfunction may be associated with the low Rs.

P1487 - HEMODYNAMICS AND EXERCISE CAPACITY AFTER CONVERSION TO TOTAL CAVOPULMONARY CONNECTION

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Background: Although several studies have shown that conversion to total cavopulmonary connection (TCPC) is a safe and effective strategy for patients with a failing Fontan, changes in hemodynamics and exercise capacity after TCPC conversion remain unknown.
Materials and Methods: Sixty-five patients underwent TCPC conversion in our institution from 1992 to 2015. Among them, 41 patients (18 males; age, 4.7-37.9 years; median age, 22.4 years; postoperative duration, 2.4-28.7 years; median postoperative duration, 16.9 years) who underwent catheter examination after TCPC conversion were retrospectively reviewed. We investigated the change in hemodynamics and exercise capacity by TCPC conversion.
Results: When comparing between before and after TCPC conversion, end-diastolic pressure (7 ± 3 vs. 9 ± 3 mmHg, p = 0.02) and cardiac index were significantly increased (2.2 ± 0.6 vs. 2.6 ± 0.5 l/min/m2, p = 0.001), pulmonary vascular resistance was significantly decreased (2.3 ± 1.3 vs. 1.6 ± 0.7 unit times m2, p = 0.003), and central venous pressure (CVP) showed no improvement (13 ± 4 vs. 13 ± 3 mmHg, p = 0.73) after the procedure. New York Heart Association (NYHA) classification was improved (I: 18, II: 22, III: 1 vs. I: 29, II: 11, III: 1). The 6-minute walking distance was significantly increased (458 ± 67 vs. 520 ± 59 m, p = 0.002). When comparing the 16 patients who underwent the cardiopulmonary exercise test after TCPC conversion with the 70 age- and sex-matched Fontan patients who had not undergone conversion, we found no significant difference in peak oxygen consumption, peak anaerobic threshold, ventilation-to-carbon dioxide production slope, and maximum O2 pulse.
Conclusions: TCPC conversion contributed to the improvement in cardiac output and exercise capacity. However, CVP did not improve, and the patients who had a TCPC conversion had no difference in exercise tolerance as compared with those who had an APC-type Fontan procedure.

P1496 - FONTAN COMPLETION – DOES IT REALLY RELIEVE CYANOSIS

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Objectives: Total cavopulmonary connection (TCPC) aims for reduction of ventricular volume overload and cyanosis. However, due to the use of fenestration and the development of venovenous collaterals, some patients remain cyanotic or desaturate during follow-up (FU).
Methods: We retrospectively studied 267 patients (median age 15 (3-57) years), who underwent TCPC at our institution between 1986 and 2015. Transcutaneous oxygen saturation (SaO2) was noted and compared longitudinally. Cyanosis was defined as SaO2 ≤ 93%.
Results: Total long-term (30 year) survival was 90% (N = 226). Total long-term mortality (N = 25) significantly correlates with cyanosis during FU (20 cyanotic vs. 5 non-cyanotic patients, p < 0.001). After TCPC, on discharge from hospital, median SaO2 of the total cohort was 94% (78-100%), 95% (80-100%) in patients without fenestration and 91% (78-98%) in patients with fenestration. In 43 patients fenestration was closed during FU. SaO2

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increased to median of 96% (88-100%) ($p < 0.01$). On last FU SaO2 was 95% (69-100%) in median, 92% (72-99%) in patients who still have an open fenestration and 96% (69-100%) in those without fenestration (including patients with closed fenestration during long-term FU). SaO2 ≤ 85% was noted in 15 patients, of whom 6 died during long-term FU. Collectively, 142 patients received catheterization during long term FU; venovenous collaterals were identified in 56 and interventionally closed in 53. A total of 37% (N = 94) were cyanotic at the time of last FU. In 47% (N = 44) this can be explained by an open fenestration, in 17% (N = 16) by venovenous collaterals. In 22% (N = 21) no reason for cyanosis could be found during catheter examination and 14% (N = 13) have not yet received catheterization.
Conclusion: Our study shows that a significant proportion of TCPC patients stay or again become cyanotic at longterm FU, with open fenestration and venovenous collaterals the most frequent reason.

P1532 - PULMONARY ARTERIOVENOUS FISTULAS IN ABERNETY SYNDROME ABOUT A CASE IN AN 18 YEAR OLD GIRL

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Introduction: "Abernethy syndrome" is a rare congenital vascular defect defined by diverting portal blood away from liver. It is often associated with multiple congenital abnormalities. We present a case of Abernethy malformation associated with diffuse pulmonary arteriovenous fistulas.
Observation: We report the case of an 18-year-old girl with dyspnea and cyanosis since the age of 6 years with progressive aggravation. Clinical examination noted the presence of conjunctival jaundice with digital hippocratism, hepatomegaly of 15 cm and arterial oxygen saturation of 65%. On the other hand, the hemodynamic constants are stable. Transthoracic Doppler echocardiography showed a left superior vena cava discharging into the right atrium with several pulmonary arteriovenous fistulas (AVF). The thoracoabdominal angioscanner confirms the presence of multiple pulmonary AVFs predominant in the bases associated with a porto-cava congenital shunt (drainage of the portal vein in the inferior vena cava) with portal hypertension and a porto-systemic Collateral venous communication. We retain the diagnosis of Abernethy's malformation. The curative treatment remains the liver transplantation to make disappear the pulmonary AVF causes of a major cyanosis.

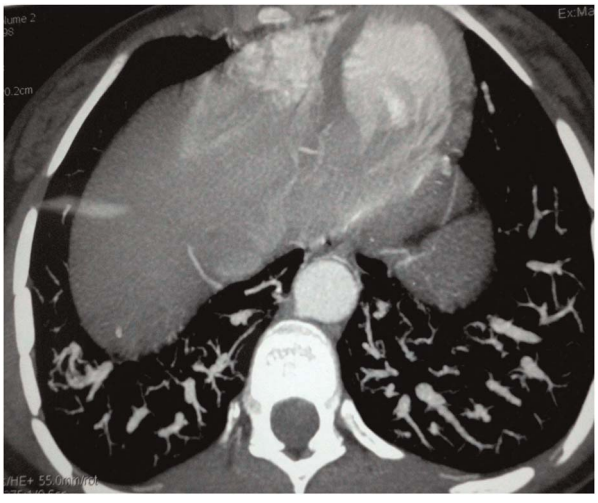

Figure 1.

P1534 - CLINICAL IMPLICATIONS OF CIRCULATING ENDOTHELIAL CELL IN ATRIAL SEPTAL DEFECT WITH PULMONARY HYPERTENSION

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Background: Patients with atrial septal defect (ASD) may stay symptom-free till adulthood. However, without treatment, pulmonary hypertension (PHN) and even Eisenmenger can be developed. Therefore, early detection of PHN and predicting its reversibility are important. We tried to define the prediction value of circulating endothelial cells in ASD with PHN.
Method: During 2014 to 2016, adult patients, older than 20 year-old, with ASD who intended to receive transcatheter closure were enrolled. All patients underwent hemodynamic study, transthorasic echocardiography (TTE) and transesophageal echocardiography (TEE). Circulating endothelial cell (CEC) and endothelial progenitor cells (EPC) numbers detection were performed using flow cytometry during cardiac catheterization. After ASD closure, they were followed for at least 6 months. Post-closure hemodynamics were evaluated either by repeating catheterization or TTE.
Result: Totally 78 ASD patients (M/F: 18/57) were enrolled; mean age: 46.6 ± 14 years-old. Among them, 27 patients (34.6%) developed PHN. 66 patients (84.6%) underwent transcatheter closure, 9 patients (11.5%) underwent surgical closure, and 3 patients (3.8%) did not receive any intervention. For the 75 patients receiving interventions, we divided them into 3 groups: Group A- initial PHN without regression, 7.6%; Group B- initial PHN with regression, 21.2%; Group C- no PHN, 71.2%. We found 74% of patients with initial PHN regressed after ASD closure. Using multivariate regression analysis, the pre-closure mean PAP was the most important predictor of PHN resolution. The CEC count was highly correlate to mean PAP and PVR. However, adding CEC counts didn't increase prediction value of PHN regression.
Conclusion: In our study, 74% of PHN regressed after closure of ASD. Pre-closure PAP is the most important predictor of the resolution of PHN. CEC numbers, though highly correlative to PAP and PVR, isn't a good predictor of resolution of PHN.

P1537 - FRAGILE AORTA IN REOPERATION CLOSURE RESIDUAL LEFT TO RIGHT SHUNT VENTRICULAR SEPTAL DEFECT IN ADULT FEMALE WITH TETRALOGY OF FALLOT

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Introduction: Reoperation post Total Correction in Tetralogy of Fallot is indicated if the patient presents deterioration hemodynamic status regarding the existing residual defects. The most disastrous complication is fragile aorta due to massive fibrosis and adhesion. Adult Tetralogy of Fallot has imbalance inflammatory process in response to traumatic factor to the cardiovascular structure due to decreased anti-inflammatory process comparing to infant and children. It results massive inflammation and fibrosis to post total correction Tetralogy of Fallot.
Purpose: To report an 18-year-old female with Tetralogy of Fallot undergoing reoperation closure residual left to right shunt VSD complicating right heart failure with fragile ascending aortic wall suspected aortitis resulting exanguination during the procedure.
Case: An 18-year-old female with Tetralogy of Fallot was hospitalized a month following total correction procedure due to

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massive right pleural effusion and pericardial effusion.The echocardiography showed large residual VSD left to right shunt and decreased right ventricle systolic function. She underwent reoperation to close residual VSD. There was massive adhesion between sternum and aorta complicating the dissection process resulting aortic rupture. Its anterior portion was extremely fragile and thin. Pericardial patch was applied to close the ruptured portion of aorta, but there were several failures in attempting to repair. The patient was death due to exanguination during procedure.
Discussion: Massive adhesion in pericardial space in post total correction in adult Tetralogy of Fallot is due to the decreased activity of anti-inflammatory agent regarding injured cardiovascular tissue. The dissection process and cannulation to aorta in reoperation results aortic injury due to the fragile aorta resulted massive inflammatory process in previously injured aorta. Femoral artery cannulation should be mandatory if there is massive adhesion in anterior aortic wall proved by preoperative thoracic CT-scan before reoperation procedure done in adult Tetralogy of Fallot.

P1538 - SINGLE INSTITUTION 20YEARS EXPERIENCE OF PULMONARY AUTOGRAFT OPERATION

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We presented our experience of Pulmonary Autograft Operation (Ross procedure) for children and adults with congenital aortic valve (AV) pathology. Ross procedure (RP) may be an alternative for mechanical and biologic valves replacement.
Objective: To examined long-term outcomes after RP for twenty-year period in our centre.
Matherial and Methods: 1996y to 2015y 200 consecutive patients have performed RP in Ukrainian Children's Cardiac Center (UCCC). The mean age on date of operation was 145 ± 101 months (from 1 to 648 months). Male - 149 (74.5%) and female – 51 (25.5%). There were 48 (24%) patients in the age younger 5 years, 36 (18%) - 6-10 years, 59 (30%) - 11-17 years and 57 (28%) older then 18 years. Aortic stenosis in 103(51.5%) patients, aortic insufficiency - 68 (34%) and mixed lesion - 29(14.5%) patients.
Results: Mortality was 18 deaths (9%), in early postoperative period - 13 deaths (6,5%), in late - 5 deaths (2.9%). Long-term results followed in 170 (93.4%) patients. Ten (5.9%) patients required reoperations for the neo-AV (4 AV repair and 6 AVR), 32 (18.8%) patients had the RV-PA conduit procedure - RVOT procedures (n = 4) and conduit replacement (n = 28), for n = 32 (18.8%) cases were performed balloon valvulotomy. Freedom from reoperation on autograft was - 98.9%, 95.4%, 93.7% after 10, 15 and 20 years. Freedom from reoperations on RV-PA conduit was 84.1%, 83.6%, 82.4 respectively. Freedom from balloon valvulotomy on RV-PA conduit was 83.7%,82.5%, 82.5% respectively.
Conclusions: UCCC have the highest experience follow-up for patients with congenital AV pathology after RP in Ukraine. Reoperations on autograft were rare 5.9%, reoperations on RV-PA conduit (18.8%). The RP is a safe procedure with low mortality.

P1539 - LONG TERM PULMONARY FUNCTION IN VENTRICULAR SEPTAL DEFECT OPERATED ADULTS – THE VENTI TRIAL

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Background: Ventricular septal defects (VSD), successfully closed within the first years of life, are considered to have great prognoses and consequently; the majority receive little or no follow-up in their adult life. However, we have recently demonstrated decreased oxygen uptake and abnormal ventilation pattern during exercise in VSD-operated adults compared with healthy controls. Whether a link to a possible physiological alteration can be found in the respiratory system remains unclear. We perform dynamic spirometry on adults, surgically treated for VSD in childhood, in order to evaluate long-term dynamic pulmonary function.
Material and Methods: Adult patients surgically treated for VSD and healthy, matched controls are included. Control subjects are matched on age and gender. Participants undergo spirometry at rest for the measurement of forced expiratory flow in one second (FEV1), forced vital capacity (FVC), and peak expiratory flow (PEF). Thirty participants will be included in both groups by March 2017.
Results: Currently, ten VSD-operated patients (23.7 ± 1.2 years) and seven control subjects (24.5 ± 1.5 years) have been included. Patients' mean age at surgery was 1.9 ± 1.7 years. No differences were found between the two groups regarding age, body mass index, body surface area or gender. Operated VSD-patients revealed lower FEV1 (3.7 ± 0.7 L) compared with healthy controls (4.9 ± 1.1), p = 0.014, as well as a reduced PEF (7.6 ± 2.8 L/min) relative to that of healthy controls (10.6 ± 2.8), p = 0.026. Furthermore, VSD-patients showed a tendency towards a poorer FVC (4.9 ± 1.1 L) compared with controls (6.1 ± 1.6), p = 0.078.
Conclusion: An impaired dynamic pulmonary function was demonstrated in adults 20 years following surgical VSD-repair. These findings may explain some of the mechanisms behind the cardiopulmonary exercise limitations previously found.

P1541 - COMPARISION OF QUALITY OF LIFE IN ADULTS WITH CONGENITAL HEART DISEASE WITH AND WITHOUT SURGICAL INTERVENTIONS

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Is the quality of life of adults with congenital heart disease worse if they had previous cardiac surgery - an Australian cohort
Background: Improved survival rates of children with congenital heart disease (CHD), has led to a greater study of the quality of life (QOL) and its determinants. Does those reaching adulthood have a reduced QOL and increased symptoms of anxiety and depression if they had undergone prior cardiac surgery compared with those who had not.
Materials and Methods: As part of a multi-centre study adults with CHD were recruited from a single community-based cardiology practice and completed self-reported questionnaires on their QOL including symptoms of anxiety and depression. An independent t-test was used to compare means between the two groups; those who had prior surgery and those who had not.
Results: 135 patients, 71 males and 64 females were recruited with a mean age of 26. 89 (65%) patients had previously undergone one or more cardiac surgical interventions. There was no significant difference in the mean QOL of patients who had previous cardiac surgery (82.6 ± 13.6) when compared with those who had not (83.6 ± 14.7), t(132) = -0.401, p = 0.689. Symptoms of anxiety and depression were also compared in the post surgical (8.6 ± 6.0) and non surgery (8.9 ± 5.2) groups. No statistically significant difference was found: t(129) = -0.203, p = 0.84. 86% of the post

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surgical group and 96% of the non-surgical group had a New York Heart Association (NYHA) functional score of 1.
Conclusions: Results from this cohort demonstrated no statistical difference in the QOL or symptoms of anxiety and depression between adults who had undergone surgical intervention when compared to those who had not. This welcome finding may have reflected the good functional capacity of both groups irrespective of the CHD abnormality and whether there was a need for surgical intervention.

P1550 - BENEFITS OF HOME BASED SUBCUTANEOUS INJECTION OF IMMUNOGLOBULIN FOR FONTAN PATIENTS WITH PROTEIN LOSING ENTEROPATHY

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Background: Protein losing enteropathy (PLE) is one of the most serious post-operative complications that has a significant adverse impact on morbidity and mortality in patients after Fontan operation. Frequent admissions for the PLE management often lead to poor quality of life (QOL) in those patients.
Materials and Methods: Our purpose was to clarify potential clinical benefits of home-based subcutaneous injection of immunoglobulin (Hizentra®, SC-IgG) for PLE Fontan patients. We introduced SC-IgG in 7 PLE Fontan patients and evaluated the influence of SC-IgG on their QOL: serial changes in plasma levels of total protein (TP; g/dl), albumin (Alb; g/dl) and immunoglobulin G (IgG; mg/dl) at early (ES; 2 weeks) and late (LS; 3~14months) after SC-IgG. We also estimated change in quality of life based on frequency of unexpected hospitalizations (number of admissions divided by follow-up [month]) and percentage of hospital stay due to the hospitalization (total hospital stay divided by follow-up period: %).
Results: During a mean follow-up of 8 months, plasma levels of TP, Alb, and IgG before, early and late after SC-IgG introduction were 5.0 ± 0.5, 3.4 ± 0.4, and 458 ± 178, 5.3 ± 0.5 ($P < 0.05$), 3.5 ± 0.4 ($P = 0.06$), and 552 ± 190 ($P = 0.18$), 5.5 ± 0.8 ($P < 0.05$), 3.7 ± 0.5 ($P = 0.07$), and 600 ± 127 ($P < 0.05$), respectively. In addition, the frequency of unexpected hospitalization decreased from 0.31 to 0.19 ($P < 0.05$) and the unexpected hospital stay decreased by 5% from 25% to 20%, respectively. No serious SC-IgG-related complications occurred.
Conclusions: Home-based management with SC-IgG significantly increased plasma levels of TP, Alb, and IgG, which accompanied by improved QOL in PLE Fontan patients. Therefore, home-based treatment with SC-IgG can be one of options to improve long-term outcome in PLE Fontan patients.

P1604 - IDENTIFICATION OF GENETIC VARIANTS AND CHROMOSOMAL ABNORMALITIES ASSOCIATED WITH EBSTEIN ANOMALY

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Background/Hypothesis: Ebstein Anomaly (EA) is an infrequent congenital heart defect (CHD) with considerable phenotypic heterogeneity in which right ventricle, tricuspid valve and electrical abnormalities prevail. Phenotypic diversity likely reflects an underlying genetic heterogeneity, which combined with studies based on small cohorts, has hindered high-confidence associations with genetic variants. Although a few chromosomal abnormalities and mutations have been linked to the disease, genetic etiologies have not been identified in most cases. Our Cardiovascular Care Center, a referral institution for CHD, has an unusually large cohort of EA patients that allows a comprehensive study of EA genetics.
Materials and Methods: We carried out a thorough phenotypic characterization of 147 EA patients, followed by unsupervised two-step cluster analysis to classify patients according to the presence or absence of comorbidities. Selected syndromic/familial cases were subjected to whole exome sequencing and/or comparative genomic hybridization. Variant filtering was accomplished using family members to identify high confidence associations with identified variants.
Results: In the cohort analysis, we identified a large proportion of syndromic (10.9%) and familial cases (11.6%). Molecular testing revealed high likelihood causative variants/abnormalities in most of the syndromic/familial cases studied. Our results suggest a novel association of EA with a rare chromosomal abnormality, the identification of a single gene in the 1p36 EA-associated region, as well as novel variants in familial cases with high likelihood of causality. Cluster analysis identified homogeneous endophenotypes that possibly reflect different underlying genetic etiologies. We are currently expanding our analysis to isolated cases.
Conclusions: Our data suggest that major causative genetic variants/chromosomal abnormalities can be found in a significant proportion of EA cases with thorough phenotypic evaluations and genome-scale molecular testing, raising the possibility of a role for genetic testing in the management of EA.

P1623 - CHARACTERISTICS AND OUTCOMES OF INFECTIVE ENDOCARDITIS IN AN ADULT CONGENITAL HEART DISEASE COHORT

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Background: The risk of infective endocarditis (IE) is known to be increased in those with congenital cardiac defects and to carry a significant mortality risk. Whilst pediatric cohorts have been well described, studies focused on IE in the adult congenital heart disease (ACHD) population are limited.
Materials and Methods: Of 4280 subjects in our ACHD database, we identified 56 patients over the age of 16 years (seen between 1991-2016) with 67 episodes of IE. We analyzed their clinical course and outcomes.
Results: Age at the time of IE was 37 (SD 13) years with a 4:1 male predominance. The most common underlying defects were bicuspid aortic valve (46%), complex two-ventricle circulations (30%) and ventricular septal defect (13%). Left sided lesions were dominant (57%) and infection involved prosthetic material in 39%. Importantly, cardiac surgery had been performed in the preceding 6 months in 12%. The most common symptoms on presentation were fever (72%) and dyspnea (21%). Blood cultures were positive in 91% (streptococcus 39%, methicillin sensitive staphylococcus aureus (MSSA) 28%, coagulase negative staphylococcus 10%,

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HACEK organisms 4%). Complication rates were high with severe valvular regurgitation in 31%, cardiac abscess in 13% and embolic phenomena in 34%, predominantly cerebral. Surgery was required in 45% of patients most commonly to replace a severely regurgitant bicuspid aortic valve. Streptococcus was causative in 50% of cases requiring surgery, with vegetations over 10mm in 40%. Death occurred during the admission in 8 patients (14%); these patients were more likely to have recurrent IE (50%, $p = 0.03$) or MSSA in blood cultures (50%, $p = 0.05$). The 12-month survival and IE recurrence rates were 85% and 9% respectively.
Conclusions: IE in ACHD patients leads to substantial morbidity and mortality and frequently requires early surgery, usually for hemodynamic reasons.

P1636 - QUALITY OF LIFE PSYCHOLOGICAL RESILIENCE PERSONALITY TRAITS AND ILLNESS PERCEPTION IN GROWN UP CONGENITAL HEART DISEASE

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Background and Objectives: There is increasing focus on quality of life (QOL) among patients with congenital heart disease. Studies suggested heterogeneity of physical and psychosocial predictors on QOL. Current study aimed to examine the relationships of QOL, psychological resilience, anxiety and depression, personality traits, illness perception, and clinical data of grown-up congenital heart disease (GUCH).
Methods: Two hundred and eight Chinese GUCH patients (male = 103) were recruited to complete the self-administered questionnaires: 36-items Short Form Health Survey version 2 (SF36), satisfaction with life scale (SWLS), Connor-Davidson Resilience Scale (RISC), Hospital Anxiety and Depression Scales (HADS), and NEO Five Factor Inventory (NEOFFI-3); and were compared to control subjects (N = 104, male = 41). GUCH patients also completed Brief Illness Perception Questionnaire (BIPQ), and their respective disease complexity, disease severity, New York Heart Association (NYHA) class and Warnes-Somerville Ability Index (WSAI) were retrieved for correlation analysis. Furthermore, multivariate regression was performed to identify the predictors of QOL measures, SWLS and SF36, respectively.
Results: GUCH patients had lower SWLS, SF36-physical component summary (SF36-PCS) and RISC (all $p < 0.001$), and greater depression ($p < 0.05$) than control subjects. Among GUCH patients, greater disease severity was associated with lower SF36-PCS and SF36-mental component summary (SF36-MCS) respectively (both $p < 0.01$). Higher WSAI was associated with lower SWLS, SF36-PCS, and SF36-MCS respectively (all $p < 0.01$). SWLS, SF36-PCS, and SF36-MCS were correlated with RISC, HADS, and illness perception respectively (all $p < 0.001$). RISC was positively correlated with positive illness perception ($p < 0.01$). RISC, several domains (Consequences, Concern) of BIPQ, and neuroticism were predictors of SWLS respectively (all $p < 0.05$). WSAI, RISC, and BIPQ (Consequences) were predictors of SF36-PCS respectively (all $p < 0.05$). Neuroticism, anxiety and depression were predictors of SF36-MCS respectively (all $p < 0.01$).
Conclusions: Reduced QOL among GUCH patients was associated with both physical and psychological attributes. Resilience and illness perception appeared having a role and might be potential targets for improving QOL in these patients.

P1681 - LEFT VENTRICULAR FIBROSIS AS A PROGNOSTIC MARKER IN PATIENTS WITH BICUSPID AORTIC VALVE DISEASE.

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Background: Bicuspid aortic valve (BAV) is associated with early aortic valve fibrosis and calcification. To what degree the left ventricle is affected by fibrosis and its prognostic value is currently unknown.
Methods: This was a retrospective, single center study evaluating all adult patients with BAV who had cardiac magnetic resonance (CMR) and followed at UCLA Medical Center from March 2002 to March 2016. CMR, transthoracic echocardiogram, and clinical data were reviewed.
Results: In total, 254 patients with BAV who had at least one CMR were evaluated, out of which 68 patients had adequate images to assess for the presence of absence of late gadolinium enhancement (LGE). Patients with prior cardiac surgery were excluded. A total of 29 patients were included in the study, out of which 18 patients did not demonstrate LGE and 11 patients had LGE. Patients with LGE were much more likely to have significant mean gradients across the AV (30.3 ± 7.2 mmHg vs 14.7 ± 3.6 mmHg, $p = 0.049$) as well as more likely to have left ventricular hypertrophy (10.3 ± 0.9 mm vs 7.6 ± 0.3 mm, $P = 0.01$). Patients with LGE were ten times more likely to need AVR in one year (55% vs 5.5%, $P = 0.0028$). Presence of LGE on CMR is associated with higher gradients and LVH while absence of LGE suggests lesser severity of disease and freedom from AVR at five years of 92.8%.
Conclusions: Evaluation of LGE by CMR as a marker of cardiac fibrosis is an additional tool of prognostic value when evaluating patients with aortic stenosis secondary to BAV. Assessment of LGE by CMR should be included in the care of patients with BAV.

P1692 - MULTIDISCIPLINARY EVALUATION OF LIVER DISEASE AFTER FONTAN PROCEDURE (FP)

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Background: The increasing long term survival after FP has brought significant liver consequences. However, there is no consensus regarding liver evaluation in these patients. The purposes of this study were (1) to evaluate the frequency of radiological signs of portal hypertension (RSPH), and hepatic nodules (HN) using ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI), (2) to measure the liver stiffness (LS) with acoustic radiation force impulse (ARFI), and (3) to compare imaging findings with clinical data.
Materials and Methods: In this Institutional Review Board approved cross-sectional study, 49 patients over 18 years of age and with at least 5 years after FP were prospectively recruited from our Cardiology Department from August 2014 to June 2016. These patients were submitted to laboratory screening of liver disorders, CT, MRI, US and ARFI directed to hepatic evaluation.
Results: Patients' mean age was 26.2 ± 7 years and the mean time interval from FP was 14.8 ± 7.1 years. RSPH were found in 3(6%),

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3(75%), and 32(66.7%) patients on US, CT and MRI, respectively. HN were detected in 3(6%), 19(39.6%), and 16(39.6%) patients on US, CT and MRI, respectively. Forty-one nodules were identified on MRI, the majority were hypervascular on arterial phase and 2 presented washout. There was no significant correlation between presence of HN or RSPH and clinical data, including interval after FP. The mean LS on ARFI was 2.2 ± 0.7 m/s. The LS values on ARFI were significantly higher in patients with hepatic nodules (2.64 ± 0.81 m/s vs 1.94 ± 0.49 m/s; $p = 0.002$). *Conclusion:* We found a high frequency of RSPH and HN in patients after FP with no correlation with cardiological or laboratory data. US did not detect almost half of RSPH and the vast majority of HN. ARFI values were significantly higher in patients with HN.

P1712 - CONGENITAL HEART SURGERY IN PATIENTS OLDER THAN 16 YEARS IN A PUBLIC PEDIATRIC INSTITUTION SCENARIO AND PROFILE OF A GROWN POPULATION

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Background: Major advances and refinement in the diagnosis and surgical treatment of congenital heart defects in the last four decades has resulted in an increasing number of adult survivors. It should be noted that congenital heart surgery is "reparative" and not curative.Many patients (p) will develop residual lesions and sequelae requiring reinterventions.
Methods and Results: From 2013 to 2015, 56 patients with a median age of 2 years (range 16.1-27) were operated with extracorporeal circulation. 35/56 were reoperations. 13% of the lesions were simple and 87% complex. Distribution by number of surgeries per year, reinterventions and type of lesion are resumed in table 1.41p had previous surgical interventions. The interventions were performed:on right ventricle outflow tract:14p,left ventricle outflow tract:7p,atrioventricular valve:7p,partial cavopulmonary connection (Glenn):1p,Fontan (extracardiac conduit):3p,reconversion to extracardiac conduit + Maze-Cox procedure: 2p, atrial septal defect closure: 4p, scimitar syndrome: 1p,unifocalization and right ventricle to pulmonary artery conduit: 1p. Median time of hospitalization in the cardiovascular critical care unit was 4 days (range: 2-21 d). Mortality rate was 1.7% (1p), and was related to RACHS score IV (p 0.0000)
Conclusions: 73% of patients had previous interventions. The congenital heart defects was moderate to severe in 87% of the cases Mortality rate was 1.7%, related to great complexity congenital heart disease with poor prognosis. The operated group conform a growing population in quantity and complexity. Most of them are followed by the pediatric cardiac team. Transference to adult public institution is challenging.

Table.

Period	Surgeries	Reoperations	Rachs1	Rachs2	Rachs3	Rachs4
2013	15	11	2	1	12	0
2014	21	13	1	1	18	1
2015	20	17	2	0	18	0

P1721 - TETRALOGY OF FALLOT 20 YEARS OF FOLLOW UP

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Background: Adult survivors with repaired Tetralogy of Fallot (TOF) are a growing population. Many patients require reinterventions during follow-up.
Methods and Results: 182 patients (p) with repaired TOF were studied from 1988 to 2016, mean age 25.3 years, median follow-up 20.2 years. Right ventriculotomy was performed in 76p(42%) and transannular patch in 73p(40%). NYHA functional class was I: 112p, II: 57p and III: 13p. Pulmonary regurgitation (PR) was moderate: 56p and severe: 40p. Complete right bundle branch block with QRS duration 160-200ms was present in 52p and > 200ms in 6p. Tachyarrhythmias were present in 27 p(15%), non-sustained ventricular tachycardia (VT): 8p , sustained VT: 7p, atrial fibrillation (AF) or flutter:11p. VT + AF: 5p. Some haemodynamic substrate was present in 66% of them. Seventynine p (43%) underwent one or more re-interventions at a mean of 11.6 years from repair: It was surgery in 56p and transcatheter intervention in 23p. In 69%, the major cause of reoperation was PR. Ten p developed infective endocarditis, 5 of them required surgery. There were 43 pregnancies in 16 women with 36 alive newborns. Overall survival at 10, 15, 25 and 35 years was: 99%, 98%, 94% and 82%, respectively. Mortality rate was 3.3%: 6p died at a mean age of 32.4 years, after a median time from surgery of 22.5 years. By univariate analysis, the mortality was associated with arrhythmia (p 0.001), QRS duration > 160 mseg (p 0.001), severe PR (p 0.004) reoperations (p 0.05) and infective endocarditis (p0.001).
Conclusions: The long-term survival of repaired TOF is excellent, with good functional class in most patients. At a mean time of 11.6 years after repair, 43% of patients underwent one or more reinterventions. Tachyarrhythmias were present in 15% of patients, 66% of them with some hemodynamic substrate. Late mortality was 3.3%, associated to arrhythmia, severe PR, QRS duration and infective endocarditis.

P1770 - LUNG FUNCTION IN ADULTS WITH PERSISTENT SMALL VENTRICULAR SEPTAL DEFECTS – THE VENTI TRIAL

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Background/Hypothesis: Ventricular septal defects (VSD) with no symptoms and Qp/Qs ratios below 1.5 are considered benign with excellent long-term prognoses. Nonetheless, we have recently demonstrated an inferior oxygen uptake during exercise among these adult VSD patients compared with healthy controls. Oxygen uptake depends on cardiac as well as pulmonary function, and whether these findings are due to pulmonary pathology needs to

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be clarified. The aim of this study was to investigate the long-term pulmonary function in adults with small persistent VSDs.
Materials and Methods: In a prospective long-term follow-up, adults with small persistent VSD's and healthy, age- and gender-matched controls between 18 and 30 years were included. Participants underwent extended lung function testing including Multiple-Breath-Washout (MBW), Impulse Oscillometry (IOS), Body Plethysmography, Dynamic Spirometry, and Diffusion Lung Capacity (DLCO).
Preliminary Results: At December 2016, 5 VSD patients (23.4 ± 4.2 years) and 8 controls (24.5 ± 1.5 years) were included. VSD patients had a lower FEV1 of 3.66 ± 0.62 L compared with 4.9 ± 1.1 L in the control group, p = 0.04. There was a tendency towards lower FRC in the VSD group (3.3 ± 0.8 L) than in the control group (4.4 ± 1.1 L), p = 0.07. There were similar tendencies between the VSD and the control group in terms of FVC (4.6 ± 1.1 L vs. 6.1 ± 1.6 L, p = 0.9), PEF (7.9 ± 2.4 L/min vs. 10.6 ± 2.3 L/min, p = 0.07) and alveolar volume (5.2 ± 1.1 L vs. 6.8 ± 1.6 L, p = 0.07), respectively. There were no differences between groups regarding sex, age, BSA, BMI.
Conclusion: The preliminary data suggests that adults with persistent VSD's have impaired dynamic lung function compared to healthy controls.

P1779 - RESULTS OF HEMODYNAMIC CORRECTION OF COMPLEX CONGENITAL HEART DEFECTS IN ADULTS

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Background: The influence of age on the results of hemodynamic correction of complex congenital heart defects remains disputable. This retrospective study is aimed to analyze immediate results of hemodynamic correction in adults.
Materials and methods: During the years 1983-2016, five hundred seven patients underwent hemodynamic correction of complex congenital heart defects (Fontan operation or bidirectional cavopulmonary anastomosis). The most frequent diagnoses were tricuspid atresia, single ventricle, and complete atrioventricular canal with pulmonary stenosis. Sixty six of 507 patients were older than 18 years of age. Out of this adult group, thirty five patients underwent bidirectional cavopulmonary anastomosis and 31 – Fontan operation. The most frequently used modification of Fontan operation was extracardiac conduit (n = 23). In 23 cases, Fontan operation was performed as a staged surgical intervention after bidirectional cavapulmonary anastomosis. Immediate results of surgical treatment were followed during the hospital period.
Results: Hospital mortality reached 9% (6/66). Lethal outcomes were caused by heart failure, hemorrhage or pulmonary thromboembolism. There were no differences in hospital mortality rate between Fontan operation and bidirectional cavopulmonary anastomosis (9.6% vs. 8.5%, p > 0.05). Different complications occurred in 21 (32%) patients. The most frequent complication was congestive heart failure and hemorrhage. Arterial blood saturation at discharge after Fontan operation and bidirectional cavopulmonary anastomosis was 92.6 ± 5.6% and 77.6 ± 6.6%, respectively (p < 0.05).
Conclusion: Hemodynamic correction of complex congenital heart defects in adults is accompanied by good immediate results and significantly improves patients' condition.

P1824 - THREE MONTHS OF RIGHT VENTRICULAR VOLUME LOADING IN RATS DOES NOT INDUCE FIBROSIS

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Background/Hypothesis: Many Tetralogy of Fallot patients are subjected to chronic right ventricular (RV) volume load after surgical repair, due to pulmonary regurgitation. The role of fibrosis in the adaptation to chronic volume loading is unknown. To determine a putative role for antifibrotic therapies in the long-term preservation of the volume-loaded RV, we aimed to establish the relation between chronic volume loading and cardiac fibrosis.
Materials and Methods: Wistar rats were subjected to aorto-caval shunt surgery (n = 24) or sham surgery (n = 15), and sacrificed after 1, 2 or 3 months. Cardiac function was measured monthly by echocardiography. Cardiac fibrosis was measured with Masson-trichrome staining. Fibrosis-related RV gene expression levels were measured using q-PCR.
Results: Effective volume loading was achieved , as cardiac output in shunt animals more than doubled compared to sham (see table). Neither cardiac output nor TAPSE changed over time, indicating adequate adaptation. RV remodeling was demonstrated by dilatation, increased ventricular and atrial weights and increased cardiomyocyte size compared to sham (p < 0,05). At all time points, Masson-trichrome stainings were not different from sham. This was validated by identical RV gene expression levels of collagens 1 and 3. Likewise, MMP2, vimentin, PCOLCE and LOXL2, markers for extracellular matrix degradation, fibroblast activation, collagen maturation and collagen crosslinking respectively, showed no differences between the groups.

Table.

	1 month		2 months		3 months	
	Sham (n = 3)	Shunt (n = 8)	Sham (n = 6)	Shunt (n = 8)	Sham (n = 6)	Shunt (n = 8)
Weights						
RV (g)	0.18 ± 0.01	0.30 ± 0.04*	0.18 ± 0.01	0.30 ± 0.05*	0.21 ± 0.04	0.39 ± 0.12*
RA (g)	0.03 ± 0.00	0.07 ± 0.01*	0.04 ± 0.01	0.07 ± 0.03*	0.04 ± 0.01	0.12 ± 0.05*
LV + IVS (g)	0.62 ± 0.03	0.91 ± 0.06*	0.66 ± 0.03	0.99 ± 0.16*	0.76 ± 0.10	1.03 ± 0.36
LA (g)	0.02 ± 0.01	0.05 ± 0.01*	0.02 ± 0.01	0.05 ± 0.02*	0.02 ± 0.01	0.09 ± 0.09
Echocardiography						
CO (mL/min)	140 ± 11	303 ± 52*	145 ± 10	281 ± 43*	131 ± 13	273 ± 54*
RVIDd (mm)	3.2 ± 0.8	4.0 ± 0.6	3.5 ± 0.3	4.3 ± 0.7*	3.6 ± 0.4	4.7 ± 0.8*
TAPSE (mm)	2.7 ± 0.0	3.9 ± 0.5*	2.7 ± 0.1	3.7 ± 0.3*	2.5 ± 0.2	3.7 ± 0.7*
RV histology						
CSSA (um^3)	299 ± 29	530 ± 189*	351 ± 54	621 ± 251*	442 ± 113	644 ± 139*
Fibrosis (%)	1.2 ± 0.1	1.0 ± 0.2	1.7 ± 0.7	2.3 ± 1.7	1.9 ± 0.9	2.3 ± 0.8

RV = right ventricle. RA = right atrium. LV = left ventricle. IVS = interventricular septum. LA = left atrium. CO = cardiac output. RVIDd = right ventricular internal diameter in diastole. TAPSE = tricuspid annular plane systolic excursion. CSSA = cardiomyocyte cross-sectional area. Fibrosis represents percentage of blue pixels on Masson-trichrome staining. Values are ± SD. Values marked with * are significantly different from sham (p < 0.05).

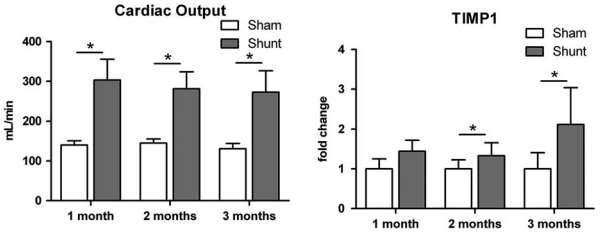

Figure 1.

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Only profibrotic marker TIMP1 was upregulated in shunt animals at 2 and 3 months compared to sham ($p < 0,05$).
Conclusions: We show that 3 months of volume loading in a rat model, equivalent to 9 years of volume loading in patients, does not result in cardiac fibrosis. It is to be elucidated whether longer duration of volume load will induce fibrosis, or that additional triggers play a role in volume load-associated fibrosis.

P1838 - THE OUTCOMES OF PREGNANCY IN WOMAN WITH CONGENITAL HEART DISEASE A NATIONAL DATABASE STUDY

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Background: For women with congenital heart disease (CHD), the hemodynamic alteration during pregnancy may increase the risks of adverse cardiovascular events. However, the population based investigation about outcomes of pregnancy in woman with CHD is still lacking.
Methods: Data on pregnancy longer than 20 weeks in woman with and without were retrieved from the National Health Insurance database records in Taiwan from 1997 to 2013. Complications and therapies were identified by their respective codes.
Results: Comparing 7933 deliveries in 5185 women with CHD and 371393 deliveries in 238833 women without CHD, the pregnant women with CHD had significant higher rates of mortality (0.11% vs 0.01%, $p < 0.0001$) and cardiac events (5.59% vs 0.59%, $p < 0.0001$). Women with CHD had higher prevalence of pregnancy-induced hypertension (3.58% vs 2.75%, $p < 0.0001$) and abortion (3.53% vs 2.73%, $p < 0.0001$). Among the cardiac events occurred in women with CHD, the leading complications were arrhythmias (3.96%), heart failure (1.37%) and stroke (0.19%). Invasive cardiac interventions during pregnancy or within 6 weeks postpartum were required in 0.42% of women with CHD.
Conclusion: Pregnancy in women with CHD are associated with significant higher rate of maternal mortality and cardiac events. Women with CHD considering pregnancy should receive careful pre-pregnant evaluations and risk stratification.

P1839 - HEALTH PROFILE IN ADULTS WITH SMALL ATRIAL SEPTAL DEFECTS

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Background: Our register based analyses of all Danish patients with a small unclosed atrial septal defect (ASD) showed a higher mortality in ASD patients with no closure compared with both the general population and with patients with a closed ASD. Furthermore, the patients without closure had an increased risk of pneumonia, atrial fibrillation and stroke.
Aim: We wish to characterize patients with small, unclosed ASDs in a prospective fashion and challenge the perception that these patients are as healthy as previously expected.
Method: In a nationwide descriptive study, the patient group consists of patients with small, unclosed ASD born before 1998. All included patients underwent TTE, ECG, 7 days Holter-monitoring, spirometry, blood samples and 6 minutes walking test. They also received questionnaires. The National Health Profile (NHP) is a self-assessment tool that allows us to compare patients with the general population ($n = 160.000$).
Results: To this point, 71 patients are included (mean age: 33 yr) and 177 excluded. In 14 patients, the defect was still open (19.7%). Aneurysmal septum was found in ten patients (14.0%). Eleven patients had mitral regurgitation (15.5%) and seven patients had aortic regurgitation (9.9%). Over one-fourth (28.2%) of ASD patients experienced high levels of stress compared with the general population (22.5%; $p = 0.07$). Most ASD patients (90%) self-assessed their health to be excellent or good, comparable to the general population (92.3%, $p = 0.66$). A minority (19.5%) self-assessed their physical function to be excellent or good. This contrasts with self-assessed physical function (37.5%; $p = 0.001$) in the general population between 25-34 yr.
Conclusion: Most defects were spontaneously closed. All valve insufficiencies were minor. Only one ASD needed catheter-based closure. Small, unclosed ASD patients more often evaluate their general health and physical function less well than the general population.

P1854 - TRANSPOSITION OF THE GREAT ARTERIES (TGA) A LATERALITY DEFECT IN THE GROUP OF HETEROTAXY SYNDROMES OR AN OUTFLOW TRACT MALFORMATION

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Background/Aim: TGA is classically considered as a so-called "conotruncal heart defect" with non-spiral great arteries. Recent studies proposed that there is probably a different mechanism involved in the genesis of TGA; it has been speculated that TGA may be a laterality defect in the group of the so-called heterotaxy syndromes (left or right isomerism) rather than an outflow tract malformation. The main aim of our study was to identify if there is indeed any association between TGA and heterotaxy syndromes, representing lateralization defects.
Methods: Restrospective cross-sectional analysis of a total of 610 patients diagnosed with TGA at King Abdulaziz Cardiac Center over a period of 13 years (2002-2015). Hospital informatics and digital data recording systems were used for collecting patients' data and reviewing echocardiograms and X-rays. All patients were reviewed to check the echocardiograms for verification of the diagnosis, type (dextro-TGA (DTGA) and levo-TGA (LTGA)) and complexity of TGA, and all other variables (such as abdominal sidedness, cardiac position, presence or absence of AVSD). Patients who did not have a proper echocardiogram, including documentation of abdominal situs, and who were repaired already in another institution were excluded (77 patients).
Results: Of 533 included patients who were verified to have TGA, 494 (92.7%) had situs solitus, 17 (3.2%) situs inversus, 11 (2.1%) left and 11 (2.1%) right isomerism. 443 (83%) of all TGA patients had DTGA. 417 (94.1%) of DTGA and 77 (85.6%) of LTGA patients had situs solitus. The incidence of right and left isomerism was found to be equal in DTGA (6 (1.4%); 5 (5.6%)) and LTGA (6 (1.4%); 5 (5.6%)), respectively. However, heterotaxy syndrome

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was more associated with LTGA than DTGA with a statistically significant difference (p = 0.004).
Conclusion: In contrary to the speculations in recent studies, our study did not reveal any significant finding supporting an association between TGA and heterotaxy syndromes.

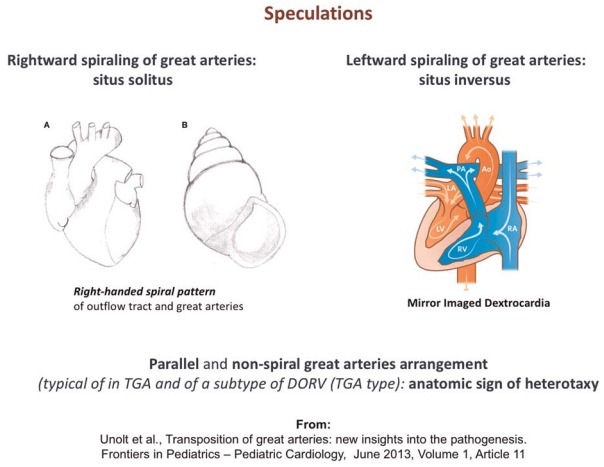

Figure 1.

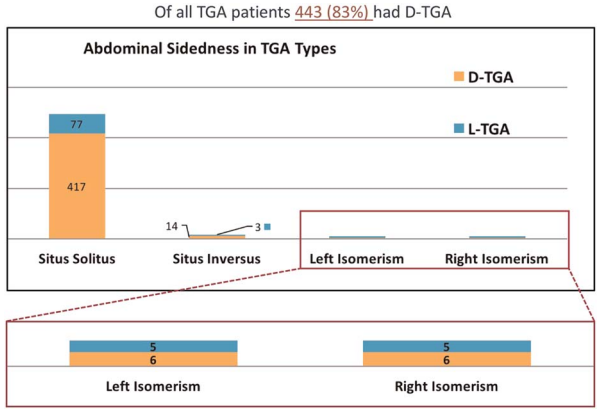

Figure 2.

P1870 - SURVIVAL RATES AND CAUSES OF DEATH IN A CONTEMPORARY ADULT CONGENITAL HEART DISEASE COHORT

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Background: Adults (age ≥16yrs) with congenital heart disease (ACHD) are at increased risk of premature mortality. There is relatively limited information concerning timing and causes of death in this population.
Materials and Methods: We studied outcomes among 2,934 adult patients (median age 33 years) consecutively seen since 2000 at our tertiary ACHD referral centre. Complete vital status and cause of death data were obtained from the Australian National Death Registry and medical records. We excluded patients with isolated patent foramen ovale or small atrial septal defects. Patients were categorised by the Bethesda classification of CHD severity (42 patients not classifiable). Those with multiple lesions were assigned to the lesion of highest complexity.
Results: By end 2016, 330 subjects (11%) had died; median age of death was 54yrs (IQR 34-72yrs) with median follow-up of 3 years. Cardiovascular death accounted for 48% of all deaths (Table 1). The leading cardiovascular causes of death were heart failure (16%), sudden cardiac death (11%), acute myocardial infarction, endocarditis and chronic ischaemic heart disease (5% each). Perioperative death was relatively uncommon at 4%. The main non-cardiac causes were malignancy (12%), pneumonia (6%) and neurological disorders (5%). Survival rate diminished as Bethesda complexity rose (p = 0.007; Table 2). Simple CHD adults had a 92% survival rate at age 65yrs. By contrast, complex CHD patients had only a 60% survival rate to age 50 years, and only 34% survived to age 65 years.
Conclusions: Our study provides contemporary insights into survival rates and causes of death in adult CHD patients seen at a tertiary referral centre.

Table 1. Percentage of cardiovascular versus non-cardiovascular death.

	Cardiovascular death (%)	Non-Cardiovascular death(%)
Simple CHD	41%	59%
Moderate CHD	46%	54%
Complex CHD	62%	38%
Total	48%	52%

Table 2. Survival table per Bethesda Classification.

% surviving at (CI 95%)	Simple CHD	Moderate CHD	Complex CHD
35 years old	99% (98-99%)	96% (94-97%)	85% (81-89%)
50 years old	98% (97-99%)	88% (84-92%)	60% (51-69%)
65 years old	92% (90-94%)	69% (61-77%)	34% (18-50%)

P1885 - SINUS OF VALSALVA ANEURYSMS A RARE CONDITION WITH A CHALLANGE DIAGNOSIS.

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Background: Aneurysm of the sinus of Valsalva is a rare congenital cardiac anomaly and its rupture can produce aortic regurgitation, cardiac tamponade, congestive heart failure, conduction abnormalities, endocarditis and stroke.
Case Report: Female, 35 years old, previously healthy. In June of 2015, she started an acute picture of heart failure (HF) characterized by dyspnea, anasarca, orthopnea and nocturnal paroxystic dyspnea. In October of 2015 she was hospitalized. The picture was interpreted as pneumonia. No improvement was obtained after treatment. An echocardiogram showed an aneurismatic image between left and right coronary sinus measuring 10mm in the major axis and severe aortic insufficiency (fig. 1). The

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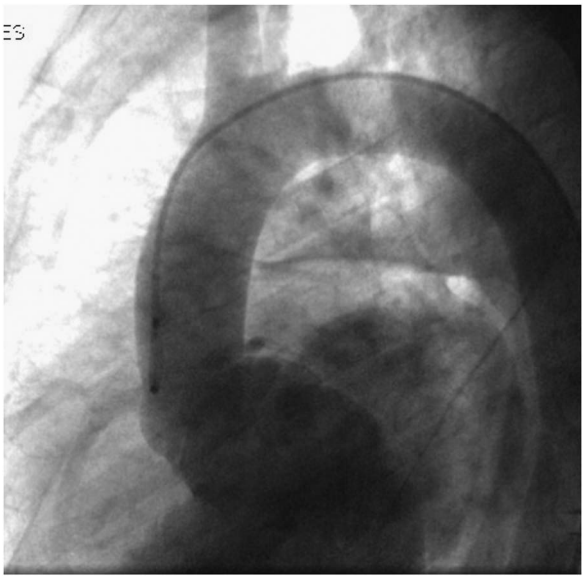
transesophageal echocardiogram confirmed the finding. The hypothesis of aortic disease was made and then she was put on furosemide and hydralazine. The patient showed slight clinical improvement and was discharged to home. After 2 months she had the first medical appointment with congenital heart disease team. Physical examination showed signs of HF such as tachycardia, orthopnea, ascites, bilateral crepitation. She was again admitted to Hospital. A hypothesis of Valsalva aneurysm (SVA) was made. MRI could not confirm due to clinical severe deterioration. A catheterization was then performed. The pulmonary capillary wedge pressure was 39 mmHg. The angiographies showed the left sinus of Valsalva aneurysm ruptured into the left ventricle (Fig 2). Soon after the procedure, the patient presented an acute myocardial infarction. She was sent to ICU and after clinical stabilization she was sent for surgery, but died during the procedure.

Conclusion: Ruptured sinuses of Valsalva aneurysms (RSVAs) may present a diagnostic dilemma because of their varied clinical presentations. In this study we reported a rare case, where no previously disease was seen and the SVA involved the left sinus.



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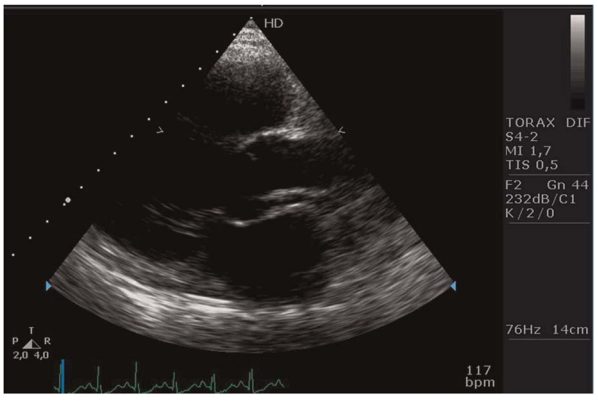

Figure 2.

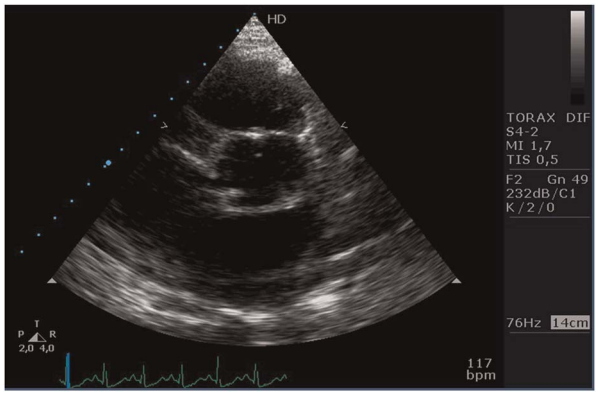

Figure 3.

P1917 - THE OUTCOME OF DEFECT CLOSURE IN SEVERE PULMONARY HYPERTENSION ASSOCIATED WITH CONGENITAL HEART DISEASE

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Objectives: This study was aimed to investigate the factors influencing clinical and hemodynamic improvement after partial defect closure in patients with severe pulmonary arterial hypertension associated with congenital heart disease.

Methods: This retrospective study enrolled patients aged more than 18 years who underwent partial closure of defects in severe pulmonary arterial hypertension (pulmonary vascular resistance >6.0 WU) associated with congenital heart disease since January 2000 to October 2016. Patient's demographics, diagnoses, functional class, hemodynamic values, and exercise capacity were investigated.

Results: Twenty eight patients underwent partial defect closure. The original diagnoses were ASD (20, 71.4%), PDA (4/28, 14.3%), VSD (3/28, 10.7%), and PDA with VSD (1/28, 3.6%). Twenty three patients showed clinical and hemodynamic improvement which was defined as improvement of functional class and decrement of mean pulmonary arterial pressure below half of mean systemic blood pressure. Seven patients discontinued pulmonary arterial hypertension-specific medications. There was no significant perioperative morbidity and mortality. The follow-up cardiac catheterizations after defect closure were performed in 14 patients, the hemodynamic values showed significant improvement ($p < 0.05$). We investigated risk factors like age at defect closure, initial functional class, initial pulmonary vascular resistance, initial pulmonary arterial pressure, initial pulmonary to systemic flow ratio, and response to vasodilators. Logistic regression analysis did not showed any significant factor that influenced on the outcome.

Conclusion: In conclusion, partial defect closure in patients with severe pulmonary hypertension associated with congenital heart disease can be a good option to improve prognosis. However careful patient selection is essential for better outcome with combined medical and surgical treatment. And close follow up is very important because pulmonary vascular resistance was still remained increased even in the patients with decreased pulmonary arterial pressure.

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Table. Improved group vs Not improved group.

	Improved	Not improved	p value
Number of patients	23	5	
Age at defect closure (yr)	35.8 (22.5 - 57.6)	36.9 (22.5 - 46.4)	0.58
Initial functional class	2.2 ± 0.7 (1 -3.5)	2.7 ± 0.3 (2.5-3)	0.16
Pre-tricuspid shunt	16 (69.6 %)	4 (80 %)	0.64
Initial Rp (Wu/m^2)	9.3 ± 2.8 (6.2- 19.3)	10 ± (7.9-12.1)	0.57
Initial PAP (mmHg)	55.5 ± 11.2 (43.0-76.0)	49.6 ± 9.8 (37.0 - 62.0)	0.30
Initial Qp/Qs	2.3 ± 1.0 (1.1 -6.5)	1.9 ± 0.3 (1.6-2.2)	0.30
Positive response to vasodilator	14 (60.9 %)	2 (40 %)	0.40

Median (lower-upper), mean ± SD, yr; year, Rp ; pulmonary resistance. mPAP ; mean pulmonary arterial pressure, Qp/Qs ; pulmonary flow/systemic flow

P1934 - USING MOCK CIRCULATION SYSTEM TO ASSESS A NOVEL EFFICIENT HANDMADE TRILEAFLET POLYTETRAFLUOROETHYLENE VALVED CONDUIT WITH SINUS

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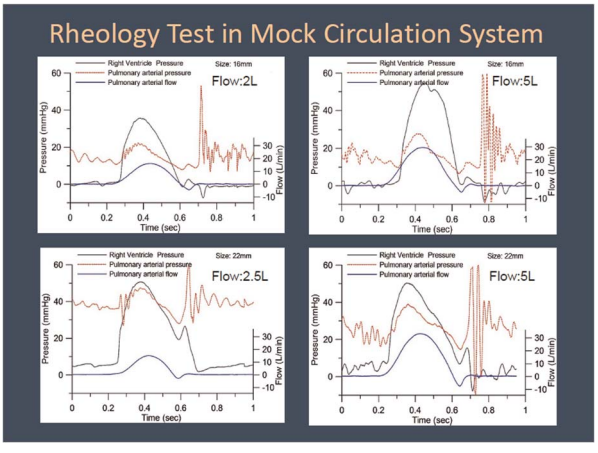
No ideal prosthetic material has been recognized for pulmonary valve replacement(PVR) and right ventricular outflow tract (RVOT) reconstruction yet. We had developed an efficient handmade trileaflet polytetrafluoroethylene (PTFE) valved conduit with sinus. The mock circulation system was used to evaluate the rheological effects of five different size conduits of 16, 18, 20, 22, and 24 mm in vitro. It showed the sinus function well. The average valve opening areas was around 70% of the cross section area in all sized conduits. The pressure gradient acrossing the valve was less than 10 mmHg in flow 5 L/min in all the five sized conduits except 16mm one which had the pressure gradient 15 mmHg in flow 2 L/min. The 16 mm valved conduit may be feasible for the patients with body weight less than 10 kg. These valved conduits have been applied to 36 patients in 7 years (2011-2016). The mean age was 15.08 ± 11.9 years (5months-49years). The early surgical mortality was 5.5%(2/36). The mean pressure gradient of the survival is 12.0 ± 12.0 mmHg under echocardiography. The PR degree less than or equal to mild in 28, mild to moderate in 5. There is no conduit explantation. The freedom from reintervention rate is 94.0%.

Conclusion: The rheology assessment results under mock circulation system examination including the pressure gradient acrossing these valve conduits and the opening areas were satisfactory for the clinical application for the patients needed PVR. The long term results have to be evaluated.



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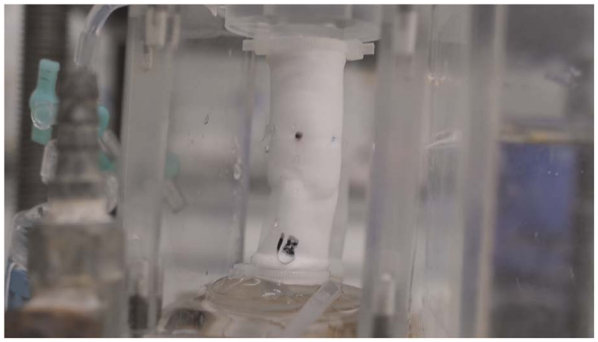

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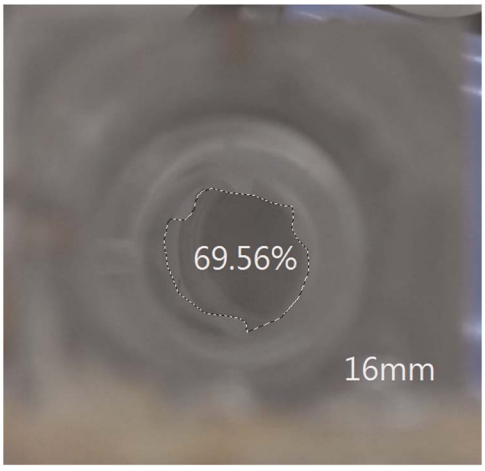

Figure 3.

P1935 - GLOBAL ADULT CONGENITAL CARDIAC NEEDS EVALUATION SYSTEMS ACCESS STUDY

Laila Akbar Ladak[1], *Disty Pearson*[2], *Kathy Jenkins*[3], *Muneer Amanullah*[4], *Kaitlin Doherty*[5], *Amy Verstappen*[2], *Malika Fatima*[6], *Babar Sultan Hasan*[6]
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Background: Adults with congenital heart disease (ACHD) have distinct health care needs. However limited data is available from lower middle income countries (LMIC). This descriptive exploratory study conducted in Pakistan aimed to assess ACHD needs from patients' and health care providers' (HCP) perspective.

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Materials and Methods: A brief e-mail survey was sent to the pediatric and adult cardiologists of five institutions (3 public) that provide ACHD services in Pakistan. An additional detailed survey was conducted at only one private institution. A telephone survey was conducted of ACHD patients. Descriptive statistics were used for data analysis.
Results: Surveyed ACHD patients (n = 128) had a mean age of 29.4 ± 10.4 years and age at surgery 26.3 ± 1.1, with a follow-up period 3.1 ± 0.7 years. 50% were male and 70% were acyanotic. The majority (75%) had a high school education and were working or housewives. Most of the surveyed ACHD (89%) patients perceived they don't need life-long care after surgery and (96%) were interested in learning about CHD, life style modifications and complications related to marriage/pregnancy. The majority of the HCPs responded that 75% CHD children would need lifelong care, yet only 10-25% return to their cardiology clinics. Cost, travelling, lack of awareness about continuity of care and unavailability of ACHD specialty were the most commonly identified barriers in the provision of care. Most HCPs (75%) were interested in learning about transition of ACHD care and management particularly during pulmonary hypertension and pregnancy. Educational sessions and support group were identified as potential resources for patients.
Conclusion: Majority of ACHD patients in Pakistan did not know that life-long follow-up was needed, which may explain why only 10-25% of these patients returned for follow-up visits after surgery. Education regarding the need for lifelong care for ACHD patients was identified as a need to alleviate the knowledge.

P1967 - CONTEMPORARY FOLLOW UP OF PATIENTS POST ATRIAL REDIRECTION SURGERY FOR TRANSPOSITION OF THE GREAT ARTERIES

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Background: A significant body of adult congenital patients born with transposition of the great arteries (TGA) have undergone Mustard or Senning procedures and require follow up. These patients often have long-term complications such as arrhythmia, baffle related problems and ventricular dysfunction. We sought to describe the outcomes of such patients at our centre.
Methods: Patients currently attending the ACHD clinic with a diagnosis of TGA who had undergone Mustard or Senning procedures were identified using the Northern Ireland regional congenital cardiac database. Data was collected regarding demographics, medications, clinical and functional status; MRI and echocardiography follow up, cardiopulmonary exercise testing and BNP. Data was analysed using IBM SPSS statistics and simple correlation performed.
Results: Forty-six patients were identified, 31 were male (67%). Mean age was 32.2 years (±6.2 years), 32 patients had undergone a Senning procedure (70%). Fourteen patients (30%) were NYHA class II or more. Nineteen patients (41.3%) were on no cardiac medications at all. Cardiac MRI demonstrated mean RVEDV of 213.4mls ±74.1mls (indexed 115 mls/m2 ±34.8 mls/m2). Mean RV ejection fraction was 53.8% ±8.1%. There was no evidence of baffle obstruction; four patients were noted to have baffle leaks. Mean BNP was 232.8 ±192.7. BNP appears to correlate with RVEF on MRI. Twenty-six patients had undergone cardiopulmonary exercise testing. VO2max achieved was 23.8 mls/kg/min ±5.7 mls/kg/min. This was 60.6% ±12.1% of estimated. NHYA class and use of cardiac medications correlate with VO2max.
Discussion: Long-term outcomes for surviving patients following atrial repair of TGA is encouraging in this group. Although RV dilatation is evident on MRI, ventricular function is appears well preserved. The majority of patients are in NYHA class I and perform well on cardiopulmonary exercise testing.

P2016 - PER CUTANEOUS CLOSURE OF RUPTURED SINUS OF VALSALVA PERILS OF THE ROAD LESS TAKEN

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Background: Sinus of Valsalva Aneurysm is a rare congenial cardiac anomaly. It is reported to be more common in the Asian population and is closely associated with the presence of a subpulmonary ventricular septal defect. Rupture of the Sinus of Valsalva is a known and potentially catastrophic complication. While medical management is directed towards stabilization, the definitive management involves closure of the ruptured aneurysm. While surgical closure remains the gold standard, in recent yeas per-cutaneous closure of the aneurysm is gaining popularity. We report two cases of per-cutaneous aneurysm closure and also a rare complication we encountered in one of the patients.
Methods: This was a retrospective case record review of 2 patients who underwent per-cutaneous closure of RSOV aneurysm in our institute between 2014-2016.
Results: The 2 adult patients presented with sub-acute onset of congestive cardiac failure. Clinical examination suggested a large left to right shunt with evidence of aortic run-off. Echocardiogram confirmed a ruptured sinus of valsalva aneurysm to right ventricle (Patient 1) and right atrium (patient 2) respectively. Both patients underwent per-cutaneous closure of the RSOV with Lifetech Ductal Occluders (size 20/18 in patient 1 and 18/16 in patient 2). The procedure would be presented in detail. 6 hours post procedure patient 1 developed acute onset pulmonary edema and left ventricular dysfunction. The device was in good position and coronary angiography was normal. Acute afterload mismatch after closure of a large shunt was suspected and intra-aortic balloon counter pulsation (IABP) was instituted. The patient recovered in the next 48 hours.

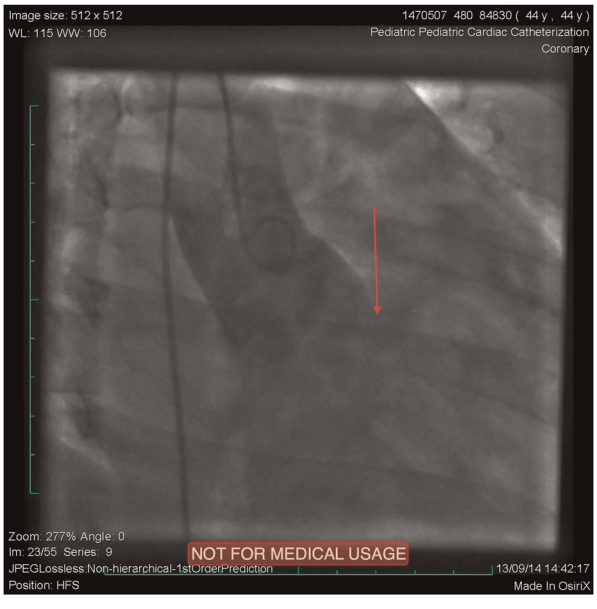

Figure 1.

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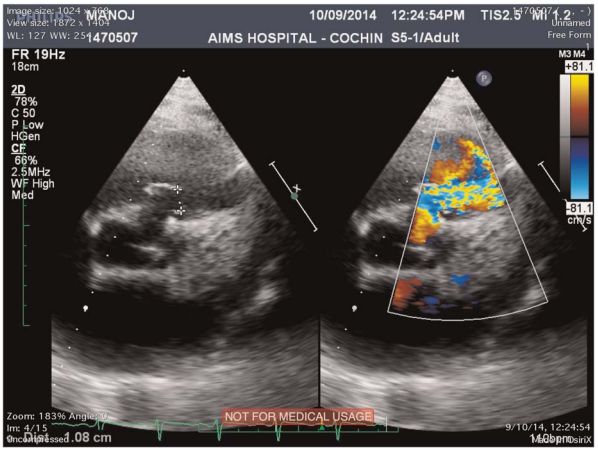
Conclusion: Afterload mismatch is a potential complication of closure of large left to right shunts. We report the first case of this occurring after per-cutaneous RSOV Closure and it's successful management,



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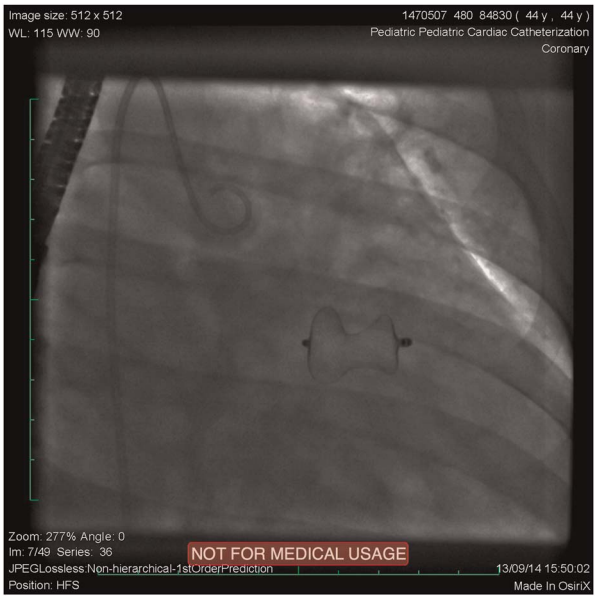

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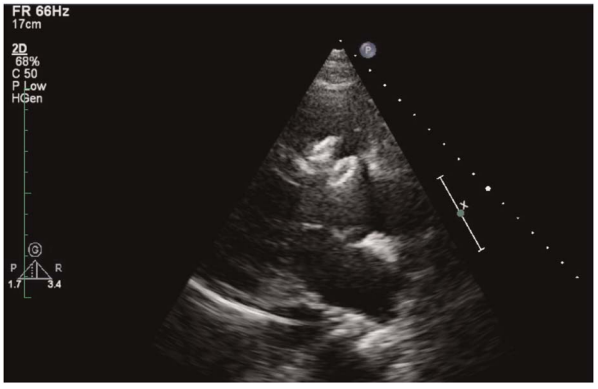

Figure 4.

P2058 - PREGNANCY IN WOMEN WITH CORRECTED AORTIC COARCTATION UTEROPLACENTAL DOPPLER FLOW AND PREGNANCY OUTCOME

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University Medical Center Groningen, University of Groningen, Department of Cardiology, Groningen-The Netherlands[1]; University Medical Center Groningen, University of Groningen, Department of Obstetrics, Groningen-The Netherlands[2]; Academic Medical Center, University of Amsterdam, Department of Obstetrics, Amsterdam-The Netherlands[3]; Academic Medical Center, University of Amsterdam, Department of Cardiology, Amsterdam-The Netherlands[4]; Erasmus Medical Center, University of Rotterdam, Department of Cardiology, Rotterdam-The Netherlands[5]; University Medical Center Utrecht, University of Utrecht, Department of Obstetrics, Utrecht-The Netherlands[6]; University Medical Center Groningen, University of Groningen, Department of Cardiothoracic Surgery, Groningen-The Netherlands[7]

Background: Women with repaired coarctation of the aorta (rCoA) are at risk of hypertensive disorders and other complications during pregnancy. Hypertensive disorders in pregnant women are associated with inadequate uteroplacental flow, which is related to adverse offspring outcome. We aimed to compare cardiac function and uteroplacental flow parameters of healthy women and women with rCoA, and investigate the relationship of maternal cardiac function, placental function and pregnancy complications in women with rCoA.
Materials and Methods: We included 49 pregnant women with rCoA and 69 healthy pregnant women from the prospective ZAHARA-studies (Zwangerschap bij Aangeboren HARtAfwijkingen, pregnancy in congenital heart disease). Clinical evaluation, echocardiography and uteroplacental Doppler flow (UDF) measurements were performed at 20 and 32 weeks gestation. Univariate regression analysis was performed.
Results: Comparison of rCoA and healthy women. In rCoA women, tricuspid annular plane systolic excursion (TAPSE) decreased during pregnancy (26.1mm to 23.4mm, $P = 0.002$). No difference in UDF indices was found between rCoA and healthy women. One woman with rCoA developed ventricular tachycardia during pregnancy. Overall obstetric and offspring events did not differ between patients and controls. However, offspring of rCoA women had a lower birth weight (3233 g vs. 3578 g, $P = 0.01$). Lower birth weight was associated with the use of β-blocker during pregnancy ($\beta = -418.0$, $P = 0.01$). Association of cardiac function and uteroplacental flow. Right ventricular (RV) function before pregnancy (TAPSE) and at 20 weeks gestation (TAPSE and RV fractional area change) were associated with impaired UDF indices (umbilical artery pulsatility index at 20 weeks $\beta = -0.02$, $P = 0.01$, resistance index at 20 and 32 weeks $\beta = -0.01$, $P = 0.02$ and $\beta = -0.02$, $P = 0.01$ and uterine artery pulsatility and resistance index at 20 weeks gestation $\beta = -0.02$, $P = 0.05$ and $\beta = -0.01$, $P = 0.02$).
Conclusions: Women with rCoA tolerate pregnancy well. However, RV function is altered (possibly through interaction of left and right ventricle) and affects placental development negatively.

P2072 - INFLAMMATORY AND NEUROHORMONAL BIOMARKERS IN ADULT PATIENTS WITH ATRIAL SEPTAL DEFECT

Anne Sif Lund Ovesen[1], Camilla Nyboe[2], Sebastian Udholm[1], Vibeke Hjortdal[1]

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Background: Atrial septal defect (ASD) is either treated conservative or by surgical/transcatheter closure. Conservative treatment is chosen for patients with hemodynamically insignificant defects or with serious comorbidity. Our previous studies showed a higher prevalence of atrial fibrillation and pneumonia in adult ASD patients with no closure compared to both the general population and to patients with a closed ASD. Both atrial fibrillation and pneumonia are associated with increased levels of inflammatory and neurohormonal biomarkers. The aim of this present study is to evaluate biomarkers in adult patients with unclosed ASD and compare them to patients with closed ASD and a healthy control group. Furthermore, the level of biomarkers will be compared to lung function, size of right atria and right ventricle.
Material and Methods: The study is conducted as a clinical case-control study. The study includes four groups of patients: 1) Spontaneously closed ASD, 2) hemodynamically insignificant ASD, 3) hemodynamically significant ASD, 4) closed ASD five years after closure. A group of healthy controls are used as reference. Plasma concentration of inflammatory and neurohormonal biomarkers are assessed, including: Interleukin (IL)-6, IL-8, tumor necrosis factor-alpha, atrial natriuretic peptide and brain natriuretic peptide. Furthermore, a spirometry is performed to examine the lung function, and the size of the shunt, the right atria and the right ventricle are measured by an echocardiography.
Results: At present 79 patients have been investigated, and data from the blood samples are pending.
Discussion/Perspectives: Results will improve the understanding of the pathological mechanisms of the complications related to atrial septal defects. A complete understanding of the physiological and hormonal changes in these patients will facilitate follow-up and might change indications for closure of atrial septal defects.

P2133 - SUCCESSFUL TOTAL CORRECTION OF MIDDLE ADULTHOOD TETRALOGY OF FALLOT PATIENTS A SERIAL CASE REPORT

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Pediatric and Congenital Heart Surgery, National Cardiovascular Centre Harapan Kita, Jakarta-Indonesia

Total correction of middle-adulthood tetralogy of fallot (TOF) patient is still controversy due to increased operative risk. Chronic hypoxemia and heart disease related comorbidities are associated with bad post-operative outcome. However, researches showed that the mortality rate of adult TOF patient who undergone total correction is 5,1%. This encourages congenital heart surgeon to optimize the management of late-presenting TOF patient rather than treat them conservatively. The authors report four TOF patients with age more than 40 years old (ranged between 43 and 53 years old) who were successfully corrected in National Cardiovascular Centre Harapan Kita. These survived TOF patients came to the hospital with low functional value and comorbidities such ischemic stroke and deep vein thrombosis. Fortunately, all of the patient have good left and right ventricle function and suitable to be operated. Intra-operatively, 3 patients were treated using valve-sparing method due to adequate size of pulmonary valve annulus. Patient were observed in intensive care unit with the average length of stay was 2,5 days and none of the patient experienced severe post-operative morbidity. Three months follow-up of the patients showed good improvement of functional value compared to the preoperative condition. The longest followup (2 years) of a patient showed no hospital re-admission related to heart problem and able to do active daily living without any marked limitation. Despite of their ages and condition, total correction is remain feasible to lengthen the life-expectancy and improve functional value of the patient.

P2143 - LATE DIAGNOSIS OF COR TRIATRIATUM CASE REPORT

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Background: Cor triatriatum is a rare congenital heart disease, characterized by the anomalous connection of the four pulmonary veins in a postero-superior accessory chamber in relation to the left atrium (LA).
Case Report: This study aims to report the case of a 20-year-old female patient with a history of cardiac murmur, as well as a complaint of dyspnea on major efforts and atypical chest pain since age 17 and an echocardiographic diagnosis of ostium secundum atrial septal defect (ASD). Due to the worsening of the symptoms, as well as episodes of syncope without prodromes, she was referred for surgical treatment. The preoperative transthoracic echocardiogram showed a membrane dividing the LA, with partial limitation to the flow of the pulmonary veins from the LA to the mitral valve, therefore establishing the diagnosis of cor triatriatum. Echocardiogram also showed an ASD with bidirectional shunt, right atrial and ventricular enlargement, right ventricular overload, and a Qp : Qs of 2.5:1. The patient underwent surgery, in which repositioning of the partial anomalous drainage of pulmonary veins, membrane section and clearing of LA was performed, as well as atrio-septoplasty with bovine patch. There were no complications in the immediate postoperative period and during the patient's hospitalization. Transthoracic echocardiography was performed after three months, showing unrestricted flow in the LA, normal cavity dimensions and normal biventricular function. After more than a year of follow-up, she is still asymptomatic.
Discussion: Despite the late diagnosis, the patient presented excellent surgical results and clinical evolution. The detailed evaluation of a patient with cardiac murmur associated with mild dyspnea is important, since adequate diagnosis of the disease may allow a curative treatment, as in the case reported.

P2161 - RIGHT VENTRICULO ARTERIAL COUPLING IN REPAIRED FALLOT PATIENTS WITH PULMONARY VALVE DYSFUNCTION BEFORE AND AFTER PULMONARY VALVE IMPLANTATION A CMR STUDY

Gianfranco Butera[1], Francesca Pluchinotta[1], Giorgia Galli[1], Luciane Piazza[1], Francesco Secchi[2], Francesco Sardanelli[2], Antonio Saracino[1], Angelo Micheletti[1], Mario Carminati[1], Massimo Lombardi[2]
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Background: Right ventricular-pulmonary arterial coupling plays an important role in the occurrence of right ventricular failure. Ventricular-arterial coupling is defined as Ea (arterial elastance)/ Emax (maximal systolic ventricular elastance). Ea/Emax can be measured using CMR as = end-systolic volume/stroke volume.

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Aim: Objective of this study was to evaluate by cardiac magnetic resonance (CMR) the right ventricular-arterial coupling before and after surgical or transcatheter pulmonary valve implantation in subjects who underwent surgery for tetralogy of Fallot and have pulmonary valve dysfunction.
Patients and Methods: We evaluated 124 patients (age 25 + /-12 years; BSA 1.23 + /-0.84) treated for tetralogy of Fallot who have pulmonary valve prevalent regurgitation, prevalent stenosis or both and underwent pulmonary valve implantation (surgical or transcatheter). CMR studies were performed before and 12 months after pulmonary valve implantation. The volumes and stroke volumes were adjusted for body surface area.
Results: Forty-nine subjects were submitted to a surgical procedure while 75 underwent a transcatheter approach. All subjects completed the pre-operative and the 12 months follow-up evaluations. Right ventricular volumes reduced significantly (RVEDD pre 109 + /-43 vs post 93 + /-23; $p = 0.001$; RVESD pre 55 + /-32 vs 44 + /-18; $p = 0.01$). Right ventricular stroke volume reduced (55 + /-19 vs 49 + /-10; $p = 0.01$) while Right ventricular ejection fraction increased (44 + /-12% vs 50 + /-11%; $p = 0.01$). The Right ventricular coupling changed significantly between the pre-operative and the post-operative period (pre 1.10 + /-0.8 versus post 1.86 + /-0.45; $p < 0.001$). Left ventricular coupling did not change. No relationships were found with age at procedure, age at evaluation, sex and group of treatment (surgery versus transcatheter). Similar changes were found when analyzing single subgroups of pulmonary valve dysfunction (prevalent regurgitation, prevalent stenosis, bith lesions).
Conclusions: In our series procedures of pulmonary valve implantation change significantly the right ventricular- pulmonary artery coupling at 12 months of follow-up.

P2204 - FONTAN CONVERSION ATTEMPTING TO PRESERVE THE SINUS NODE IN ANTI ARRYTHMIA SURGERY

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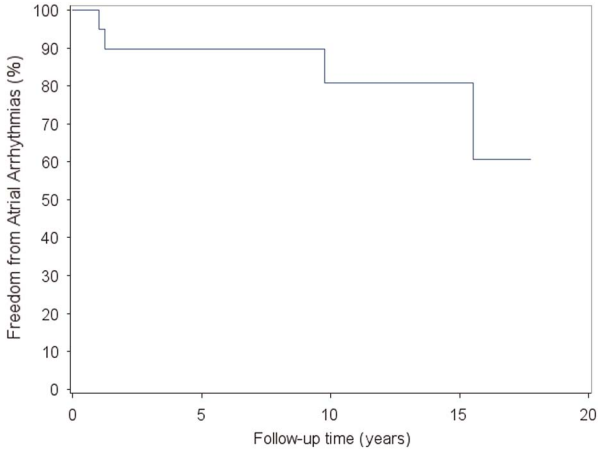
Starship Hospital, Green Lane Paediatric and Congenital Cardiac Services, Auckland-New Zealand

Objectives: To ascertain if an alternative limited right atrial maze (RAM) technique during Fontan Conversion preserves sinoatrial node function, chronotropic reserve and atrioventricular conduction with low incidence of pacemaker dependency.
Methods: A retrospective cohort study of consecutive patients undergoing Fontan Conversions with anti-arrhythmia surgery at a congenital cardiac surgical unit between 1997 and 2015. Of the 22 patients identified, 19 had the alternative limited RAM technique that aims to preserve sinus node function. Mortality, preservation of atrioventricular conduction, pacing requirement, and recurrence of atrial arrhythmia were reviewed.
Results: The median age at Fontan conversion was 28 years and the median interval from original Fontan surgery was 19 years. Concomitant anti-arrhythmia surgery was performed in all patients. Indications included atrial arrhythmias (91%) and exertional fatigue (73%), with a median follow up of 11 years (range 1 to 19 yrs). 19 patients are known to be alive, 2 of whom are lost to regular cardiology follow-up. There was one early post-operative death (4.5%) and 2 patients have subsequently died at 11 months and 7 years post conversion (overall mortality 14%). A pacing unit was placed in 16 (73%) after surgery one having been paced pre-operatively. 4/22 (18%) are pacemaker dependent for intra-atrial block (1) or atrioventricular block (3, with congenitally corrected transposition) with 50% of patients retaining chronotropic competency as assessed by Holter or exercise testing. Four had recurrence of atrial arrhythmia (18%) and in 2 the recurrence occurred >5 years after conversion.
Conclusions: Fontan conversion with an alternative limited right atrial maze technique can be undertaken with an acceptable mortality risk. Long term follow up demonstrates an enduring benefit from the conversion surgery in the majority of patients with inhibition of atrial arrhythmia recurrence and a low incidence of pacemaker dependence compared with standard anti-arrhythmia surgical techniques.



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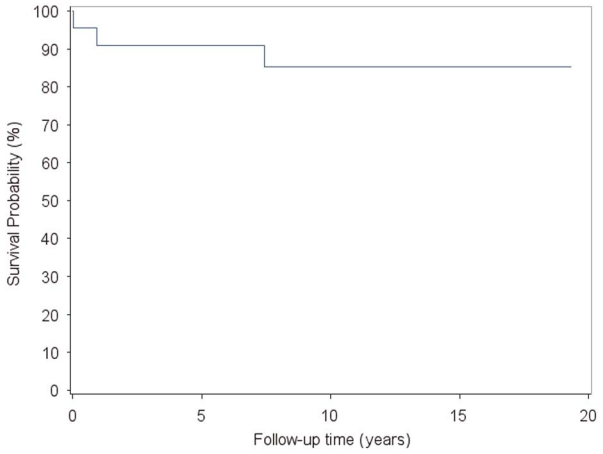

Figure 2.

P2213 - CORONARY ARTERY MICRO FISTULAS ARE COMMON COMPLICATION IN POLYSPLENIA WITH SINGLE VENTRICULAR PHYSIOLOGY

Hiroki Mori[1], Hisashi Sugiyama, Saeko Yoshizawa[2], In-Sam Park[1]
Tokyo Women's Medical University, Pediatric Cardiology, Tokyo-Japan[1]; Tokyo Women's Medical University, Pathophysiology, Tokyo-Japan[2]

Introduction: Coronary artery fistula (CAF) is a rare anomaly in patients with congenital heart diseases. In clinical, coronary artery micro fistula related to congenital heart disease could be demonstrated in patients with polysplenia.
Hypothesis: Coronary artery micro fistulas could be not rare anomaly in patients with polysplenia and cause myocardial damage.

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Methods: Total 62 patients (male/female: 24/38, age: 23 ± 1.1 years) with polysplenia, who underwent cardiac catheterization, were retrospectively reviewed. Of those 46 had single ventricle physiology. Furthermore, 2 pathological specimens were evaluated to detect myocardium damage and coronary artery micro fistulas.
Results: Coronary angiography showed coronary artery micro fistula in 11 patients (18%). All had single ventricular physiology. Proximal potion of coronary artery, which supply micro fistula, run with tortuous and dilation. All coronary artery micro fistulas terminated into right ventricle chamber (Figure A). Statistical difference was not observed in oxygen saturation and end-diastolic pressure of systemic ventricle in these patients compared with the patient without fistulas. Pathological examination showed multiple fistula-like vessels in endocardial myocardium (Figure B; arrow head, scale bar 1000 μm). The vessel diameter ranged from 300 μm to 1000 μm. Those had irregular vessel wall with immature smooth muscles, elastic fiber and collagen fiber (Figure C and D; scale bar 100 μm). Furthermore, remarkable perivascular interstitial fibrosis was observed in the right ventricle.
Conclusions: The prevalence of coronary artery micro fistula could be clarified as 18% in patients with polysplenia with single ventricular physiology. Furthermore, coronary artery micro fistulas was proved on pathological specimens. Perivascular interstitial fibrosis in the right ventricle could be caused by coronary micro fistula.

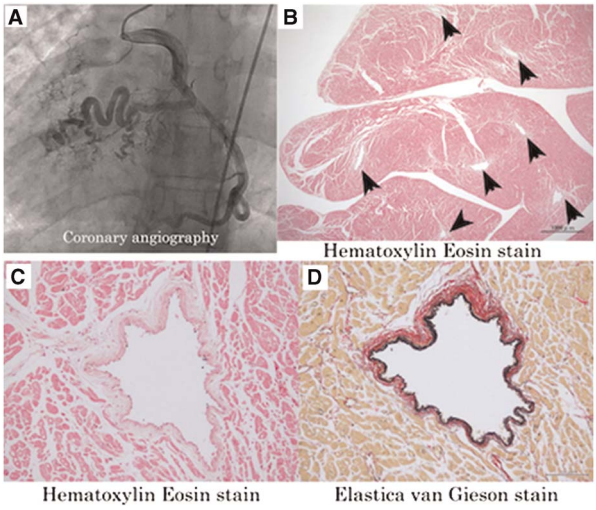

Figure 1.

P2219 - FONTAN CONVERSION FUNCTIONAL OUTCOMES OVER TIME – A SINGLE CENTRE EXPERIENCE

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Objectives: To evaluate the functional impact of Fontan Conversion surgery.
Methods: A retrospective cohort study of consecutive patients undergoing Fontan Conversion with antiarrhythmia surgery at a congenital cardiac surgical unit between 1997 and 2015. Twenty two patients were identified. Survival was 86.5% with one perioperative death and two later deaths. Two patients have been lost to followup. New York Heart Association (NYHA) class, ventricular function assessment, cardiac catheter and exercise testing were evaluated.
Results: NYHA Class improved by at least 1 grade in 79% of survivors. At 10 year follow up (FU) 10 of 13 patients are in NYHA class I-II. Two patients have subsequently managed successful pregnancies. Ventricular function has deteriorated in 4 patients (18.2%) after 7-15 years FU with 2 of these having severe dysfunction, whilst 5 (22.7%) have ongoing improved function after 4-19 years FU. Cardiac catheter haemodynamics were available prior to and a median of 6 years after conversion in 8 patients. Seven of these had improvement in at least one parameter of pulmonary artery pressure, pulmonary arterial wedge pressure, pulmonary vascular resistance or cardiac output. One patient required neo-left pulmonary artery stenting post Fontan conversion. Eight patients have had cardiopulmonary exercise testing pre and post conversion with a median of 7.5 years between (range 1-13 years). Pre conversion the median age-predicted peak VO2 was 47% (range 44-64%), and post the median was 58.5% (range 45-72%). Ten patients (45%) have not required antiarrythmic agents since surgery, whilst 4 are being treated for atrial arrhythmia recurrence and 4 for non-sustained ventricular arrhythmia. Two patients have permanent anti-tachycardic atrial pacing.
Conclusion: Fontan conversion at our centre has provided improved functional capacity in the majority with a longitudinal retention of benefit. Careful patient selection, timing and techniques to address arrhythmias will optimise results from Fontan

P2223 - THE EFFICACY AND SAFETY OF DIRECT ORAL ANTICOAGULANT IN ADULT PATIENTS WITH FONTAN CIRCULATION

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Background: Thromboembolic complications occurring after the Fontan procedure are well documented. Up to now, how to prevent these events remains unclear. On the other hand, Direct Oral Anticoagulants (DOAC) has been used to prevent thromboemboli events for the patients with atrial fibrillation and venous thromboembolism (VTE).
Objective: To investigate the safety and efficacy of DOAC in Fontan patients.
Methods: Sixty patients after Fontan procedure (27 men, 27 ± 8 years old, mean duration after the procedure is 19 ± 7 years, 13 patients with Atriopulmonary connection and 47 with Total cavopulmonary connection) were reviewed retrospectively. Patients, prescribed DOAC, warfarin or antiplatelet, were classified into 3 groups. Cases having no prophylactic therapy were excluded. The clinical outcomes were analyzed. Primary endpoints were thromboembolic complications and bleeding events. Thromboembolic complications were defined as symptomatic cerebral infarction, peripheral arterial embolism, and VTE. Bleeding events were hemorrhage requiring medical intervention including cessation or change of the anti-thromboembolic medication and/or more than 2 g/dl or 10% decrease in hemoglobin.
Results: Twenty-five cases were classified as DOAC group, 17 cases in antiplatelet group. Warfarin were used in 15 cases. On the other hand, three cases were having no prophylactic therapy. Mean duration of the medication were 29 ± 14, 55 ± 36, 67 ± 50 month, respectively. No case had thromboembolic event. But two patients in each of DOAC and Warfarin group had bleeding event. No case had bleeding event in Antiplatelet group.

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These showed no significant difference among three groups regarding estimated event free survival rate.
Conclusion: Direct oral anticoagulants in adult patients with Fontan Circulation may be safe and efficacious as observed in warfarin or acetylsalicylic acid.

P2235 - LV NON COMPACTION IN PATIENTS WITH SINGLE VENTRICLE HEART DISEASE IS ASSOCIATED WITH IMPAIRED SYSTOLIC FUNCTION

Preeti Choudhary[1], Christian Hamilton-craig[2], Shelby Kutty[3], Raj Puranik[4], David Celermajer[4]
University of Sydney, Department of Medicine, Sydney-Australia[1]; The Prince Charles Hospital, Department of Cardiology, Brisbane-Australia[2]; University of Nebraska Medical Centre, Department of Cardiology, Nebraska-United States[3]; Royal Prince Alfred Hospital, Department of Cardiology, Sydney-Australia[4]

Background & Aims: LV non-compaction is a congenital morphologic abnormality of the LV myocardium, postulated to occur due to early embryonic arrest of myocardial compaction and is associated with adverse outcomes including cardiomyopathy and premature death. Although observed in association with various congenital heart lesions, its effect on cardiac function remains unknown. We sought to assess the prevalence of LVNC in patients with univentricular physiology (SVHD) and its effects on ventricular function.
Methods: Cardiac magnetic resonance imaging of 93 patients with SVHD (mean age 24 + /-8 years; 55% male) from 3 tertiary congenital centres were retrospectively reviewed. The presence of LVNC was defined as having a non-compacted: compacted (NC:C) myocardial thickness ratio >2.3:1. The distribution of LVNC, ventricular volumes and function were correlated with clinical data.
Results: The prevalence of LVNC was 37% with a mean of 4 + /-2 affected segments. The distribution of LVNC was apical in 100%, midventricular in 29% and basal in 17% of patients. Patients with LVNC had significantly higher end-diastolic (128 + /- 44 vs. 104 + /-46 mL/m2, p = 0.047) and end-systolic LV volumes (74 + /-35 vs. 56 + /-35 mL/m2, p = 0.039) with lower LV ejection fraction (44 + /-11 vs. 50 + /-9%, p = 0.039) compared to those without LVNC. Stroke volumes did not significantly differ in the two groups (65.0 + /-29 vs. 73 + /-37 mL /m2, p = 0.36). The number of segments involved did not correlate with adverse ventricular function (p = 0.71).
Conclusions: LVNC is common in patients with SVHD physiology with predominantly apical and mid-ventricular involvement. Although the LVNC distribution is not extensive, its presence is associated with increased indexed ventricular volumes and impaired global systolic function.

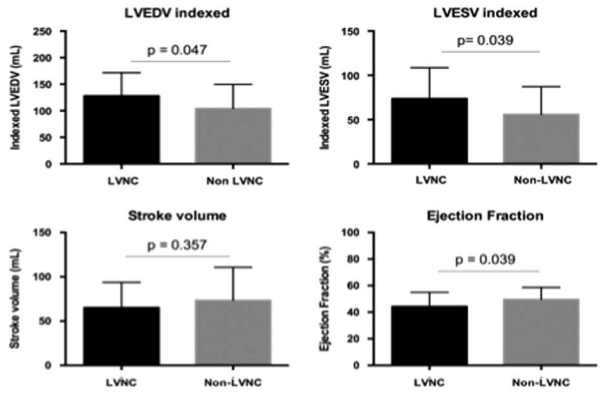

Figure 1.

P2248 - DOUBLE WHAMMY A RARE CASE OF PACEMAKER LEAD THROMBUS AND PULMONARY EMBOLISM IN A PATIENT WITH CCTGA AND ESSENTIAL THROMBOCYTOSIS

Chukwuemeka Oguguo, Jaspal Dua, Petra Jenkins, Heiko Schneider, Ashish Shah, Andreas Hoschtitzky
Manchester Heart Centre, Cardiology, Manchester-United Kingdom

A 73year-old male with congenitally corrected transposition of great arteries (ccTGA), severely dilated/impaired systemic RV/Pulmonary embolism (PE), presented with worsening LV function and pacemaker lead thrombus with PE, refractory to heparinization/warfarin.
Background: The pacemaker was inserted 12 years prior for complete heart block (CHB), without undergoing transthoracic echocardiogram (TTE) when he first presented, delaying ccTGA diagnosis. He had suffered a PE 2 years prior.
Management: 2 weeks of IV Heparin showed no resolution of clot burden therefore he was taken for surgical resection. Through aorto-bicaval cardiopulmonary-bypass, opened resection of large organised thrombi in RA/RV associated with RA/RV pacing leads, pacemaker removal and permanent epicardial dual-chamber pacemaker implantation. Due to post-operative thrombocytosis, he was referred to haematologists, who diagnosed with essential thrombocytosis with a rare mutation, increasing his thromboembolic risk.
⋆Evidence of large clot on TOE on 4CH view
⋆Further evidence of large clot on lead on TTE
- Note filling defects in right pulmonary tree, indicative of pulmonary embolism
Discussion: ccTGA - uncommon form of congenital heart disease (CHD); incidence 0.02-0.07/1000 live births (<1% of CHD), characterized by atrioventricular/ventriculoarterial discordance, and rhythm disorders, most commonly CHB, with 2% increased annual risk, or progressive AV block, seen in over 1/5th of unoperated ccTGA, and can present later in life with CHB. Patients presenting de novo with CHB should undergo TTE to exclude ccTGA. Thromboembolic events can be life-threatening and atypical thromboembolism risk factors include PPM/intracardiac shunts/rhythm disorders/younger age. These risk factors need appropriate management with surveillance investigations including anticoagulation/sinus rhythm maintenance
Summary: ccTGA is rare and complex to manage. Patients have increased thrombo-embolic risk due to dilated systemic ventricles/pacemaker leads/sub-optimal treatment and in this case, compounded by haematological disorders like thrombocytosis. Low threshold for investigations/anticoagulation is important.

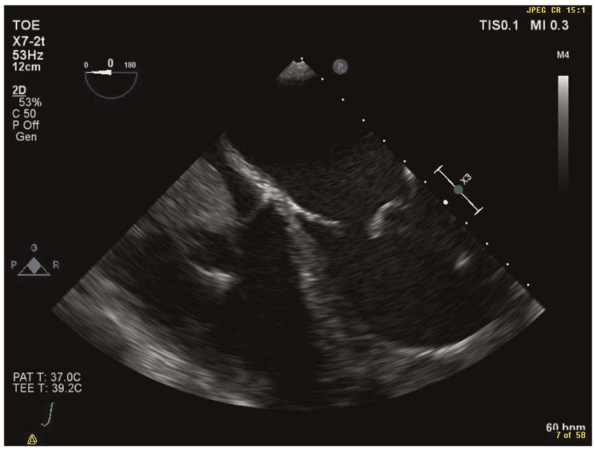

Figure 1.

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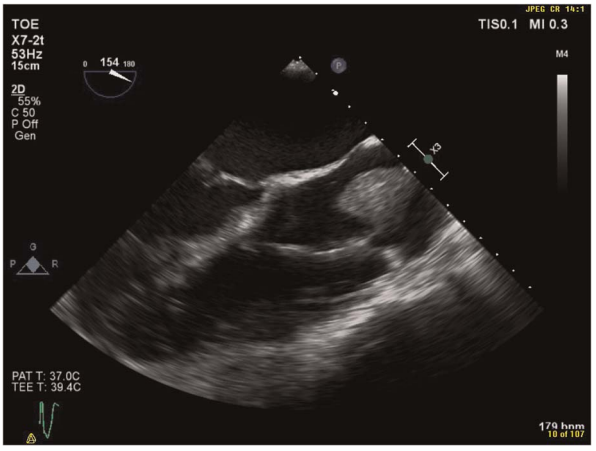

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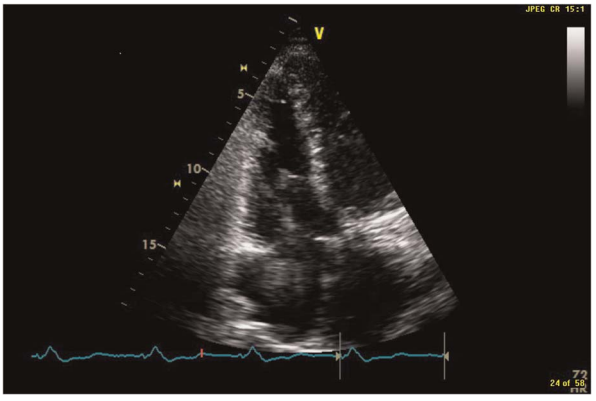

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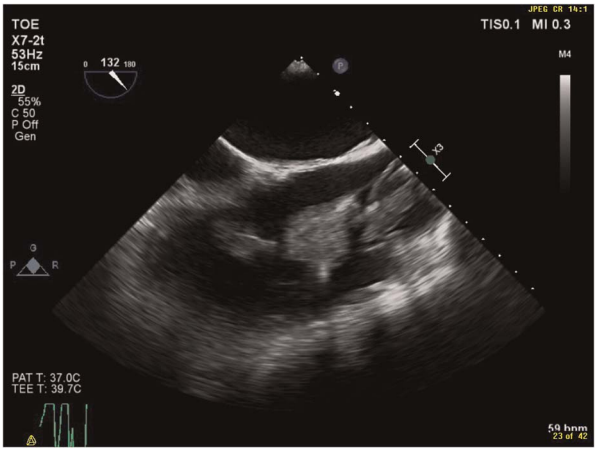


Figure 4.

P2250 - REFRACTORY ATRIAL ARRHYTHMIAS IN ATRIO PULMONARY FONTAN THE FAILING OF THE FONTAN

Chukwuemeka Oguguo, Jaspal Dua, Petra Jenkins, Andreas Hoschtitzky, Ashish Shah, Heiko Schneider, Bernard Clarke, Sahrkaw Muhyaldeen
Manchester Heart Centre, Cardiology, Manchester-United Kingdom

A 36year-old male, with tricuspid atresia, normally related great arteries, VSD and rudimentary RV, presented with recurrent atrial arrhythmias, post atria-pulmonary (A-P) Fontan procedure. His first Fontan procedure was in 1980 with a modified A-P Fontan procedure in 1998. He had developed severe systemic ventricular dysfunction.

*4CH view showing small, atrialized RV, enlarged RA

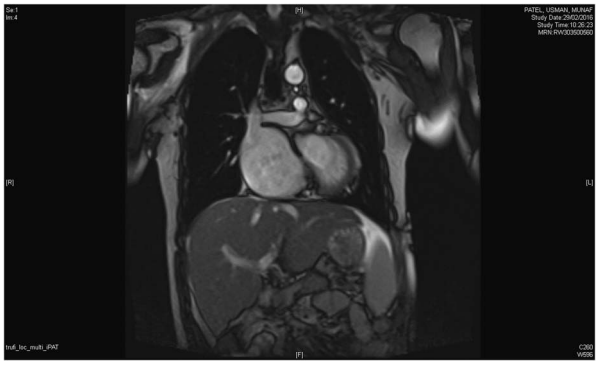
*CMR – coronal view - note grossly enlarged RA and VSD, as well as Fontan connection

From 2012, he developed various types of atrial arrhythmias including atrial flutter/AV-nodal-re-entry-tachycardia, refractory to medication, presenting to hospital, frequently. He was unsuccessfully treated on admissions with direct-current-cardioversions (DCCV) and he underwent three electrophysiology studies/ radiofrequency ablations via CARTO® system.

*CARTOablation views showing areas of ablation (red dots) thereby isolating PVs

Due to these refractory arrhythmias despite treatment, he was referred for heart transplantation, currently on the transplant list.

Discussion: 10-years post-Fontan operation, arrhythmias occur in over 50% and 20% of patients have SVTs. Intra-atrial-re-entry tachycardia/ atrial flutter, being most common. Patients present with palpitations, haemodynamic deterioration/compromise, clot formation/embolic events. Fontan arrhythmias are resistant to medication due to re-entrant mechanisms, facilitated by atrial substrates adjacent to valves/patches/suture lines, reduced systemic ventricle function and cellular injury from hypoxia/atrial stretch. Patients usually require advanced/multiple EP procedures, however these usually only modify the substrate. Nonetheless, there are good success rates with catheter-based treatment, provided there is adequate access to myocardium/cardiac chambers. Newer techniques have been introduced with increased success rates like non-invasive mapping/remote magnetic navigation. Either Fontan conversion (guided by surgical protocols), Ventricular-assist



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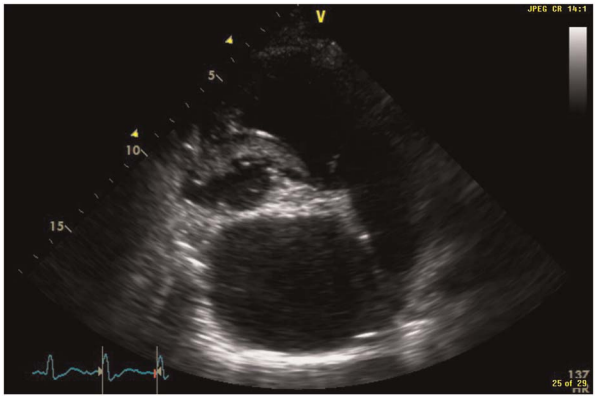

Figure 2.

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devices/cardiac transplantations are considered in refractory arrhythmias despite medical/device therapy.
Summary: Fontan arrhythmias are common, varied, problematic, life-threatening. Patients may undergo multiple DCCV/ablations, if refractory to medication. Potential need for further surgery/ catheter interventions/ transplantation should be considered, if indicated, and discussed with patient/family.

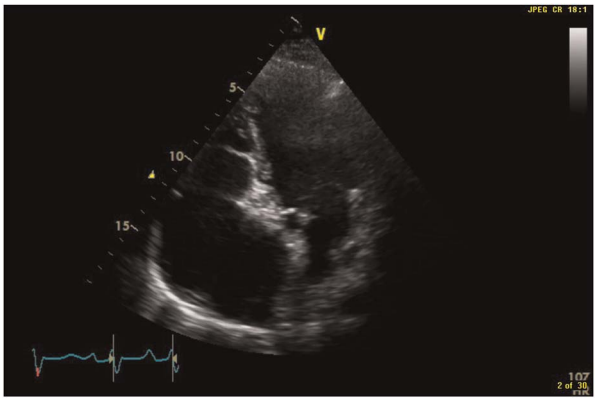

Figure 3.

P2253 - THE IMPACT OF HEART DISEASE FUNCTIONAL STATUS ON THE LIFE OF ADULT PATIENTS WITH CONGENITAL HEART DISEASE

Junko Enomoto[1], Yoshiko Mizuno[2], Shigeru Tateno[2], Koichiro Niwa[3]
Toyo University, Department of Education, Tokyo-Japan[1]; Chiba Cerebral and Cardiovascular Center, Department of Adult Congenital Heart Disease and Pediatrics, Chiba-Japan[2]; St Luke's International Hospital, Cardiovascular Center, Tokyo-Japan[3]

Background: The quality of life (QOL) and mental health of adult patients with congenital heart disease (ACHD) may be affected by their current heart disease functional status. The aim of this study is to explore the impact of heart disease functional status on their life and the strategy to improve their life.
Material and Method: A total of 199 ACHD patients and 158 reference participants (aged 20-60; no students included) completed the QOL questionnaire using a linear analog scale, the satisfaction with life scale (SWLS) and the hospital anxiety and depression scale (HADS). Patient characteristics included sex, age, educational level, marital status, employment status and subjective patient-reported New York Heart Association (NYHA) functional class. The study focused on four groups: 1) reference, 2) NYHA I, 3) NYHA II, 4) NYHA III/IV.
Results: Compared to the reference and NYHA I groups, NYHA III/IV have lower QOL and satisfaction with life, and higher anxiety and depressive symptoms. NYHA II and NYHA III/IV have patients with lower educational levels and more patients that are single. NYHA II and NYHA III/IV have fewer patients that work full time and NYHA III/IV has more unemployed patients. NYHA II and NYHA III/IV patients who work have a higher QOL and SWLS than unemployed patents.
Conclusion: While the reference and NYHA I groups have an acceptable QOL and mental health feedback, NYHA II and NYHA III/IV experience a worse situation. According to our results, if NYHA II and NYHA III/IV patients engaged in any type of work, their QOL and mental health would definitely improve. In Japan, currently, the government offers substantial financial support to ACHD patients. However, the government needs to provide more ways to help these patients become contributing members of society.

P2292 - MATERNAL AND FETAL OUTCOMES IN PATIENTS WITH POST DOUBLE SWITCH OPERATION FOR CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

Terunobu Fukuda[1], Naoto Kawamatsu[1], Yasufumi Kijima[1], Yumi Shiina[1], Shigeru Tateno[2], Koichiro Niwa[1]
Cardiovascular Medicine, St Lukes International Hospital, Tokyo-Japan[1]; Adult Congenital Heart Disease and Pediatrics, Chiba Cardiovascular Center, Chiba-Japan[2]

Background: The double switch operation (DSO) has emerged as a way to improve prognosis for the congenitally corrected transposition of the great arteries (ccTGA) with deteriorating systemic right ventricle since mid-1980's, therefore, a number of patients after DSO have been growing up to be reproductive age. However, there are paucity of reports on the pregnancy and neonatal outcomes of this condition.
Methods and Results: To clarify the course of pregnancy and neonate outcomes in patients with ccTGA after DSO, the medical records in two-specialized centers from Jan 2000 to Des 2015 were retrospectively reviewed. Four patients aged 27 ± 3.5 at delivery with preserved bi-ventricular funcntion, all of them underwent Mustard and Rastelli operation, were included. Reported pre-pregnancy problems including moderate intra-conduit stenosis with previous catheter ablation for supra-ventricular tachycardia in one, atrial tachycardia in two, and small baffle leakage in one, however, they were all in good physical conditions. Three pregnancies developed atrial tachycardia during pregnancy, one of whom required cardioversion and taking beta-blocker. One delivered 1245 g neonate at 29 week 5 day for premature rupture of the membranes. Other three women delivered at scheduled day without any maternal cardiac complications, one delivered by forceps delivery with 1984g neonate at 37 week 4 day, one underwent scheduled caesarean section with 2032 g neonate at 36 week 3 day and one underwent emergency caesarean section due to arrested labor with 2288 g neonate at 37 week 4 day.
Conclusions: ccTGA patients after DSO could complete pregnancy, if they had good bi-ventricular function and physical activity before pregnancy. However, there were frequent episodes of atrial tachyarrhythmia and had a tendency to deliver low-birth weight infants. Careful management with multidisciplinary team including cardiologist, obstetrics, anesthesiologist and other multidisciplinary teams should be required to complete these complicated cases.

P2296 - GROWN UPS CONGENITAL HEART SECUNDUM ATRIAL SEPTAL DEFECT WITH MITRAL VALVE REGURGITATION LONG TERM OUTCOME OF MITRAL VALVE REPAIR

S Laura Mazalan[1], J Jeswant Dillon[1], Intan Fariza Gaafar[2]
National Heart Institute, Department of Cardiothoracic Surgery, Kuala Lumpur-Malaysia[1]; National Heart Institute, Research Department, Kuala Lumpur-Malaysia[2]

Background and Hypothesis: Mitral valve regurgitation (MR) in Secundum Atrial Septal Defect (ASD) often attributed to clefts leaflets. In selected Grown-ups Congenital Heart groups, prolonged ASD shunts exposes the mitral valve to a more complex valve pathology with a more challenging valve repair techniques. Study objective to review our long-term experience in surgical valve repair of MR in ASD, determining valve key pathological features, complex valve repair techniques with long-term outcomes.
Material and Methods: 24 years (1992-2016), retrospective analysis of 59 patients in Grown Ups Congenital Heart Disease

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(age range: 16-65 years, mean 32.8years) with Secundum ASD and predominantly mitral regurgitation (MR) underwent surgical correction of ASD with autologous pericardial patch with concurrent mitral valve repair.
Results: 37 female and 22 male; predominantly mitral regurgitation (MR) 53(89.8%), mitral stenosis (MS) 3(5.1%) and mixed MR/MS 3(5.1%). Co-morbidities of pulmonary hypertension 21 (35.6%), atrial fibrillation 14(23.7%), endocarditis 1(1.7%). Valve predominant features were mitral leaflets prolapse 43(72%), thickening 24(40.7%), with annular dilatation 29(49.2%) and chordal elongations 16(27.1%). Prolapsed leaflets repaired with chordal replacement 14(23.7%) and shortening 7(11.8%) and annuloplasty ring placement 52(88.1%). Cleft repaired in 11(18.6%) patients. Concurrently 5(8.5%) required MAZE procedure and 26(44%) tricuspid annuloplasty repair. Mean follow-up duration of 78.1 ± 68.6 months. Above 10 years follow-up, mitral valve competency grade; fully competent 61%, trivial 20.3%, mild 16.9%, severe 1.7%. At 15 years follow-up, freedom form re-operation were 97.3%. Subsequent improvement in symptoms from pre to post-operatively in NHYA functional class I 29(49.1%) to 52(88.1%) and reversion to sinus rhythm 45(76.3%) to 48(81.4%)
Conclusions: Prolapsed mitral leaflets and annular dilatation are predominant features of prolonged uncorrected ASD shunts onto mitral leaflets. Cleft repair alone is insufficient. Chordal procedure and ring annuloplasty increase the feasibility and durability of repair.

P2297 - EVALUATING THE LONG TERM EFFECTS OF THE FONTAN PROCEDURE ON THE HEPATIC SYSTEM

Petra Jenkins[1], Zenib Zadar[2], Jaspal Dua[1], Vaikom Mahadevan[3]
CMFT, ACHD, Manchester-United Kingdom[1]; University of Manchester, Manchester Medical School, Manchester-United Kingdom[2]; UCSF, ACHD, San Francisco-United States[3]

A palliative procedure performed in univentricular congenital cardiac lesions, the Fontan procedure, is associated with impaired liver function and liver fibrosis. Currently these patients are monitored using liver function tests (LFTs) and liver ultrasound (US) scans, however these tests are targeted for viral mediated fibrosis. Acoustic radiation force impulse (ARFI) imaging measures tissue elasticity and may have an important role in assessing liver stiffness. We aimed to assess the efficacy of LFTs, liver USS and ARFI imaging in diagnosing liver fibrosis in Fontan patients. We also aimed to assess any relation between cardiopulmonary exercise test (CPET) variables and ARFI scores. Data was collected retrospectively from the Manchester ACHD Centre. We identified 12 patients who had results available for LFTs, USS Liver, ARFI and CPET. The sensitivity of LFTs and liver USS was found to be 6.6% and 86% respectively. ARFI identified liver congestion/fibrosis in all patients with 33% at F2, 33% at F3 and 33% at F4. There was found to be a slight reduction in % peak VO2 (62.6 vs 46, p = 0.2) and % predicted O2 (101.1 vs 88.9, p = 0.6) in F3 patients, with a higher VE/VO2 (32.5 and 42.0, p = 0.2), indicating a relationship in worsening cardiopulmonary variables, and hence Fontan haemodynamics and the presence of the degree of liver fibrosis.
In conclusion, LFTs alone are not a suitable screening test for Fontan associated liver disease (FALD) and liver USS does not accurately quantify the degree of fibrosis. ARFI requires further research in larger study samples to determine a role in routine hepatic monitoring of Fontan patients. Although we found a weak relationship between impaired CPET variables and greater ARFI scores, this relationship requires further investigation.

P2313 - RV FUNCTION IS IMPAIRED IN ADULTS WITH TETRALOGY OF FALLOT

Ramakrishnan Sivasubramanian, Saurabh Gupta, Ritesh Gupta, Rajnish Juneja, Shyam Kothari, Anita Saxena, Vinay Bahl
AIIMS, Cardiology, New Delhi-India

Introduction: In developing countries, quite a few patients with Tetralogy of Fallot's (TOF) present late, and sometime in adulthood. Long standing cyanosis and progressive right ventricular (RV) hypertrophy and fibrosis may have deleterious consequences on RV function. We studied the differences in clinical, echocardiographic (including speckle tracking) and angiographic parameters among TOF patients presenting late in adolescence/adulthood as compared to children with TOF.
Methods: 20 children of age ≤3 yrs (mean age 1.95 ± 0.76; range 1 3 years) and 10 patients of age ≥ 13 yrs (mean age 20.5 ± 16.5; range 13 38 years) were recruited in the study. Clinical, echocardiographic and angiographic parameters were compared between the groups. The echocardiographic parameters of RV function evaluated in this study included: end-diastolic RV diameter, RV/LV ratio, tricuspid annular plane systolic excursion (TAPSE), percentage of RV fractional area change (RV FAC), and peak systolic longitudinal strain derived from speckle tracking. Angiographically, the diameter of DTA, RPA and LPA were measured in all patients and were indexed to body surface area (BSA). McGoon ratio was also calculated.
Results: Patients with uncorrected TOF were more symptomatic as compared to children with higher functional class and lower saturation. Parameters of RV functions were relatively lower than normal in both children as well as adult with TOF. However, all the parameters of RV function were significantly more impaired in adults with TOF. For instance, TAPSE was 16.8 ± 1.5 in children as compared to 13.9 ± 1.6 in adolescents with TOF (p - <0.01). Mcgoon ratio was not significantly different.
Conclusion: Patients with uncorrected TOF's presenting in late childhood or adulthood are more symptomatic. RV dysfunction as assessed by the various echocardiographic parameters is more common in adolescents and adults than in children with TOF, which may compromise the operative outcomes.

P2325 - PULMONARY VALVE REPLACEMENT WITH AN INJECTABLE STENTED PULMONIC VALVE 10 YEARS OF EXPERIENCE

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Niguarda Hospital, Cardiovascular, Milan-Italy

Background: Chronic pulmonary valve regurgitation (PR) following repair of congenital heart diseases, such as tetralogy of Fallot, is a common late sequela often requiring reoperation. Long-standing chronic PR can result in right ventricle (RV) dilatation and failure and increased late morbidity and mortality. A timely reoperation may lead to a complete RV recovery. The conventional approach is pulmonary valve replacement (PVR) on cardiopulmonary bypass (CPB) often with cardioplegic arrest. The aim of this multicenter study is to evaluate mid to long-term outcomes of patients undergoing a less invasive PVR with the BioIntegral® Injectable Stented BioPulmonic prosthesis.
Material and Methods: From 2007 to 2017, forty-nine symptomatic patients (mean age: 28.7 ± 16.3 years, range: 10-71.4) with severe PR underwent PVR with an injectable stented pulmonic prosthesis at 13 centers. The mean interval between primary correction and PVR was 19.8 years (range: 1-45.4). Mean follow-up was 44.8 months (range: 3 months-10 years). Twenty-six patients

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(53%) underwent isolated PVR; concomitant procedures performed on CPB in twenty-three patients (47%) included tricuspid valve repair, ablation of cardiac arrhythmias, ventricular septal defect repair, pulmonary artery plasty, aortic procedure and right ventricular outflow tract enlargement. The median prosthesis size was 27 mm (range: 19-31 mm).
Results: Off-pump pulmonary valve implantation was performed in 39% of patients (19/49). Overall mortality rate was 4% not device-related. No major complications occurred postoperatively and no prostheses were explanted. Echocardiography and cardiac magnetic resonance in follow-up detected a significant reduction in RV size ($p < 0.001$) and low transvalvular gradients across PV ($p < 0.05$) (Fig. 1).
Conclusions: Injectable Stented Pulmonic valve implantation is a good alternative to conventional approach mostly in patients requiring multiple cardiac operations. The prosthesis, available up to large sizes, presents a laminar flow and no-risk of coronary arteries compression. PVR can be performed off-pump with lower biological impact, shorter implantation time and minimal morbidity. Results concerning prosthesis durability are encouraging. Mid and long-term haemodynamic performances are excellent.

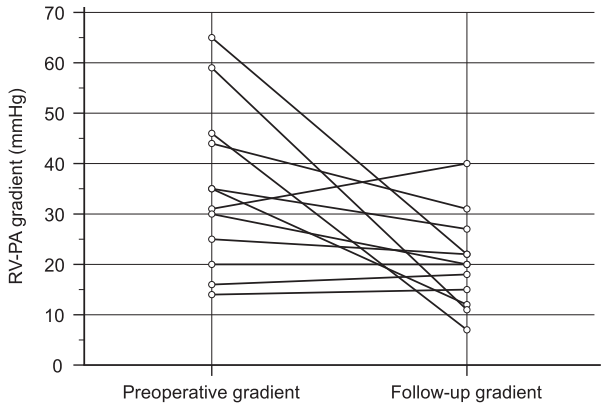

Figure 1.

P2327 - FUNCTIONAL STATUS AND INTRACARDIAC ANATOMY OF DOUBLE INLET LEFT VENTRICLE SURVIVORS WITHOUT FONTAN CIRCULATION

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Background: Double inlet left ventricle (DILV) patients are currently palliated with the Fontan procedure. Data on long-term survival with a Fontan circulation remain compromised. A small, selected subgroup of "survivors" of DILV, however, has been reported to live to a comparably high age without a Fontan procedure. The aim of this study was to determine the natural course of uncorrected DILV and describe the intracardiac anatomy.
Material and Methods: In this retrospective study, clinical data and the intracardiac anatomy of DILV patients without Fontan procedures from all Dutch congenital heart disease referral centers were analyzed.
Results: Twenty-four patients were included, of whom 16 are still alive (median age 44 years, range 12-68). Seventeen (71%) patients had pulmonary stenosis, of which 7 (44%) had a pulmonary banding procedure. Six (39%) had Eisenmenger's syndrome. The ventriculo-arterial connection was discordant in 18 (75%) and concordant in 3 (12.5%) hearts. NYHA functional class was I or II in 71% of the patients. Kaplan Meier analysis showed survival to start dropping linearly from 49 years of age, while the oldest patient was alive at 68 years. Median survival did not differ significantly between patients with and without pulmonary stenosis (62 and 53 years respectively). Mean overall transcutaneous oxygen saturation at rest was 86% (SD ± 7.2) and was significantly higher in patients with pulmonary stenosis than in patients without (88% ± 6.3 and 79% ± 5.2 respectively).
Conclusions: The data on DILV natural survivors give evidence that it is possible for a selected subgroup of these patients to live up to a relatively high age without Fontan procedures. Although the majority of these patients had pulmonary stenosis, patients with no pulmonary vascular protection who developed Eisenmenger's syndrome had equal life expectancy. Pulmonary banding might be an effective surgical alternative for certain DILV patients.

P2331 - RESULTS OF SURGICAL CORRECTION OF EBSTEIN ANOMALY OF THE TRICUSPID VALVE IN ADULT PATIENTS

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Background: Operation of Ebstein anomaly technically belongs to the demanding interventions in cardiac surgery, especially if the surgeon aim is a complex repair of the valve and therefore the results of these operations of the adults vary greatly according to the experience of each surgeon or center. Our goal is to evaluate own results in adult patients with Ebstein anomaly.
Methods: A retrospective assessment of 45 patients who underwent totally 49 surgical interventions of Ebstein anomaly between 12/2005 and 12/2016.
Results: Mean age at surgery was 33.9 ± 12.1 (19-63) years. Morphologically, most patients (72%) had Ebstein Anomaly type C. Repair of tricuspid valve was performed in 44.4% cases, bioprosthetic valve replacement in 53.3% cases. The procedures were associated with the bidirectional cavo-pulmonary anastomosis in 26.0%, mitral valve repair in 8.9%, and closure of atrial septal defect in 64.0% and right atrial MAZE procedure in 40%. The average duration of extracorporeal circulation was 147 ± 28 min with an aortic cross-clamp time of 112 ± 31 min. Two patients died within 30-days after surgery and another 2 during further follow-up. 6 patients required re-replacement of bio-prosthetic valve (2 for structural valve deformation, 22 and 11 years after first surgery, 4 for infective endocarditis in ± 4 years after first replacement). Presence of NYHA class II and more was reduced from

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2.6 ± 0.67 before to 1.58 ± 0.57 after surgery, p < 0.0001 Grade of tricuspid regurgitation after valve repair decreased from mean 3.8 ± 0.49 to 1.39 ± 0.94 after surgery, p < 0.0001.
Conclusion: In a specialized adult congenital heart disease center surgical either repair or replacement of Ebstein anomaly can be performed with acceptable mortality in adulthood. Both tricuspid valve function and functional capacity is significantly improved.

P2390 - MENSTRUAL PROBLEMS IN WOMEN WITH CONGENITAL HEART DISEASE

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Objectives: To investigate the age at menarche and the prevalence of menstrual disorders, and determine the relationship between menstrual abnormalities and cardiac conditions in women with congenital heart disease (CHD).
Materials and Methods: Women aged 16 to 60 years old with CHD who admitted our institute between February 2016 and December 2016 were enrolled this study. Patients were asked about menstruations including menarche, cycle and duration of menstruation. We analyzed the relations between menstruation and the hemodynamics, New York Heart Associate (NYHA) classification, cyanosis, and medications.
Results: A total of 121 patients were enrolled (median age 34 years, range 16-58 years). Patients were categorized according to the degree of severity of the underlying heart defect. The average NYHA class was 1.5 ± 0.6 and the average oxygen saturation rest was 94 ± 7% (range 70 – 99%) . The average age at menarche was slightly delayed in women with CHD compared with the average age in Japanese women (12.9 years vs 12.6 years). In the patients after Fontan palliation (n = 44), the age at menarche was significantly delayed compared with patients after biventricular repair (13.4 years vs 12.7 years, p = 0.002). There were 4 patients (3%) of primary amenorrhea, 27 patients (22%) of secondary amenorrhea. Overall, menstrual disorders including cycle disturbance and abnormal duration of menstruation were observed in 56 (46%) patients. The occurrence of these disorders depended on the presence of heart failure, the severity of CHD.
Conclusions: Menstrual disorders were frequently observed in patients with heart failure and complex heart disease.

P2405 - RIGHT BUNDLE BRANCH BLOCK AS A PREDICTOR OF FAVOURABLE OUTCOME AFTER PERCUTANEOUS PULMONARY VALVE IMPLANTATION

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Introduction: Recent data showed a right ventricular dyssynchrony in patients with tetralogy of Fallot (TOF). Percutaneous pulmonary valve implantation (PPVI) has become an important procedure to treat a dysfunction of the right ventricular outflow tract in these patients. Despite providing good results, there is still a considerable number of non-responders to PPVI. The authors speculated that electrical dysfunction of the right ventricle plays an underestimated role in the outcome of patients after PPVI. This study aimed to investigate the influence of right ventricular electrical dysfunction i.e. right bundle branch block (RBBB) on the RV reverse remodeling after PPVI in terms of change in indexed right ventricular enddiastolic volume (RVEDV) and right ventricular ejection fraction (RVEDV) measured by cardiac MRI.
Methods: The study included patients after correction of TOF with or without RBBB, who had received a PPVI previously at the Heart Center of the University of Leipzig, Germany in the period from 2012 to 2015. Cardiac MRI and echocardiography were performed directly before and six months to one year after the procedure in all patients.
Results: 24 patients were included. Patients without RBBB i.e. narrow QRS <120 ms had significantly better RVEF (p = 0.002) and had smaller indexed RVEDV prior to intervention compared to patients with RBBB and QRS ≥ 120 ms. Patients with pre-PPVI QRS < 140 ms showed after PPVI statistically significant difference in grade of reverse remodeling of the right ventricle with decreasing RV volumes (RVEDV pre-PPVI 132.6 ± 38.0 ms vs post-PPVI 105.4 ± 25.9 ms [mean ± SD], p = 0.002). The parameters of LV function and volume remained unaffected by RBBB. RV remodeling was only seen in patients with a QRS Duration < 140 ms.
Conclusion: The presented data indicate that the QRS duration seems to be a valuable parameter in the prediction of right ventricular reverse remodeling after PPVI, as it represents both electrical and mechanical function of the right ventricle.

P2429 - PATIENT SATISFACTION WITH AN ADULT CONGENITAL HEART DISEASE OUTPATIENT SERVICE RESULTS OF A RETROSPECTIVE SURVEY OF 178 PATIENTS

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Introduction: In 2016 the NHS England Congenital Heart Disease Review set out standards of care for Adult Congenital Heart Disease (ACHD) services including Domain 4 – ensuring patients have a positive experience of care. This is a vital part of facilitating and maintaining a long term therapeutic relationship with patients who have a chronic illness. It also highlighted the importance of key holistic elements such as provision of information to patients and their families to facilitate a collaborative approach to care, good communication and ease of access to services.
Method: A questionnaire was created comprising 25 questions and a free comment box. The survey was distributed to 178 patients attending an Adult Congenital Heart Disease clinic in a 'hub and spoke' model at Liverpool Heart and Chest Hospital between February 2012 and December 2015. Questions were detailed to assess key aspects of service provision and hoped to explore: Ease of access; Facilities provided; Collaborative care; Patient education and Overall patient experience.
Results: Results showed the ACHD service was very well received with average score of at least 9.7/10. One area of improvement was the availability of the ACHD specialist nurse (only 62.3% saw specialist nurse during their clinic visit) though when this service was available it was valued very highly in terms of courtesy and communication. Another was the provision of information surrounding ease of access to clinic. Only 82% were given information on how to get to the hospital, 62.6% were given information on public transport and 68.4% were given information on parking. All free text comments were positive in nature with common themes including feeling 'reassured' and finding staff 'helpful'.
Conclusion: Overall, this survey showed that the clinic was performing well in its capacity as an important point of contact between patients with long term illness and their healthcare providers.

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Table 1.

Question	Yes	No	Unsure	Percentage
1. Were you given information on how to get to the hospital?	146/178	27	5	82
2. Were you given information about parking?	121/177	45	11	68.4
3. Were you given information about public transport?	109/174	50	15	62.6
4. Was it easy to find the outpatient department?	172/177	4	1	97.2
8. Did the doctor listen carefully to what you had to say?	174/175	0	1	99.4
9. Were you given enough time to discuss your condition?	175/175	0	0	100
10. Did the doctor explain your heart condition?	173/175	1	1	98.9
11. Were some tests done on the day?	171/178	7	0	96.1
12. Did the doctor explain about these?	165/174	8	1	94.8
13. Did you have confidence and trust in the doctor?	176/177	1	0	99.4
14. Did the ACHD specialist nurse explain about the service?	107/108	1	0	99.1
15. Did she give you enough time to discuss your condition?	107/108	1	0	99.1
16. Did she listen carefully to what you had to say?	108/109	1	0	99.1

Table 2.

Question	Average Score
17. The courtesy of the ECHO physiologist	9.7
18. The courtesy of the clinic nurse	9.8
19. The courtesy of the doctor	9.8
20. The courtesy of the ACHD specialist nurse	9.8
21. How useful was your visit today?	9.6
22. Do you have more idea and understanding of your heart condition, after today's visit?	9.6
23. Did you feel you were involved as much as you wanted to be in the decisions about your care and treatment?	9.8
24. Overall how would you rate the doctor in terms of knowledge/friendliness and courtesy?	9.9
25. Overall how would you rate today's visit to the ACHD clinic?	9.8

P2527 - OUTCOME OF COMPRESSION OF LEFT CORONARY ARTERY IN PATIENTS WITH SEVERE PULMONARY ARTERIAL HYPERTENSION ASSOCIATED WITH CONGENITAL HEART DISEASES

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Background: Long-standing pulmonary arterial hypertension (PAH) sometimes causes the dilation of pulmonary artery (PA) associated with compression of left main coronary artery(LCA). We explored the outcome related to the risk of sudden cardiac death in patients with PAH.
Method: PAH patients with PA enlargement (≥40 mm) by echocardiography were studied with CT scanning, and had been reviewed with medical records retrospectively.
Results: Total 51 patients (mean age 49 ± 13, 33.3% male) were enrolled. No patient reported exertional chest pain or aggravated dyspnea at the time of study. However, complications associated with PA dilatation were found in 18 patients (35.1%): LCA compression were in 18 of 51(35.1%), almost in patients with CHD (16 of total 18 patients with LCA compression); airway compression, 4 of 51; PA dissection, 1 of 51 ; and thrombus in PA, 4 of 51. Patients with complications had more dilated main PA than patients without complication. The area under the receiver operating characteristic (ROC) curve for main PA diameter measured at LCA-oblique view was 0.878 (95% confidence interval (CI): 0.756 to 1.000, $p < 0.001$) with the highest discriminating sensitivity and specificity being 86.7% and 67.7%, respectively at main PA diameter of 45mm. Of total 16 patients with CHD shown LCA compression in CT, only 4 patients revealed critical compression of LCA in coronary angiography. One patient with severe LCA compression underwent invasive coronary angiography and received angioplasty with stent placement at the os of main LCA. In the others, open heart surgery for angioplasty of PA reduction had done successfully.
Discussion: The complications of PA enlargement related to long-standing PAH was high. Evaluation of LCA using CT scanning should be considered in PAH patients with dilated PA to prevent sudden cardiac death in regular follow-up.

P2532-40 YEAR FOLLOW UP OF PATIENTS WITH CRITICAL AORTIC STENOSIS CROSS SECTION IMAGING AND FUNCTIONAL STATUS

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Background: Survival with congenital critical aortic valve stenosis (CAS) can be successfully achieved by surgical or catheter interventions. Long term outcome however remains uncertain.
Methods and Results: We performed a 40 year (1970-2010) review of 96 patients, requiring intervention for neonatal (<30 days) CAS. Surgery was undertaken in 61 patients and balloon dilation in 35 patients. Follow-up data was available for 88 patients. There were 29 (32.9%) reported deaths, of which 19 (19.8%) <90 days after intervention. Overall survival rate at 1, 5, 10, 15 and 20 years was 73%, 71%, 71%, 66%, and 66% respectively. Of the 67 survivors, 53 (79%) had at least one re-intervention and 21 (31%) had more than one re-intervention. Twenty-five patients were further assessed in our centre. Median age was 15.7 ± 6.4 years. Mean peak VO2 was 32.1 ± 8.2 ml/kg/min, peak VO2 was abnormal in all but one patient (96%) and severely depressed in 13 patients (56%). Mean left ventricle (LV) ejection fraction was 65.5 ±11.22% and mean LV end-diastolic volume was

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79.96 ±17 ml/m2, Z score 0.02 ±1.4. Mean LV outflow tract Vmax was 2.27 ±1.17 m/s. Four patients (16%) had moderate aortic regurgitation. Five patients (20%) had severe LV diastolic dysfunction measured echocardiographically and confirmed by invasive measurement. Mean right ventricular outflow gradient was 19.23 ± 23.57 mmHg. Focal endocardial fibroelastosis (EFE) identified on echocardiography (n = 13, 52%) correlated with focal EFE assessed by MRI (r = 0.61, p = 0.009) but not with late gadolinium enhancement (r = 0.453, p = 0.0677). EFE was associated with early Ross intervention (r = -0.52, p = 0.0324) and diastolic LV dysfunction (r = -0.48, p = 0.0164).
Conclusions: Long-term follow-up of patients with CAS showed a high rate of mortality and re-interventions. Reduced exercise capacity, left ventricle diastolic dysfunction and EFE remain a concern long-term after initial procedure. EFE is associated with poor outcome.

P2537 - A NEW CHALLENGE FOR ADULTS WITH CONGENITAL HEART DISEASE THE PSYCHOSOCIAL APPROACH IN CARDIAC REHABILITATION

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Background/Hypothesis: Currently there are no precise guidelines about cardiac rehabilitation for adults with congenital heart disease (ACHD) even though there are indications about the possible benefits from their participation to such programs.
Materials: ACHD attending a rehabilitation program were evaluated when it comes to the psychosocial aspects, relevant to cardiovascular risk. This included biographic and lifestyle data, health perception (EuroQol) quality of life, anxiety and depression (Hospital Anxiety and Depression Scale HADS). The medical variables considered were: Six Minute Walk Test (6MWT) (at hospitilisation and discharge), Ejection Fraction, NYHA Index and diagnostic category.
Methods: The questionnaires were administered to 22 patients (16 females, age range 17-65, average age 41,36) at admittance and the EuroQol and HADS were also re-administered at discharge.
Results: An indication of improvement in health perception (EuroQol) was observed (average 54,74 SD 16,11 to 85,9 SD 13,43). Anxiety and depression levels at admittance were mostly in the normal range and they correlated strongly with one another (r = 0,63; p < 0,01). There was an improvement in anxiety in 81% and in depression in 67% of the cases. Belonging to the severe condition group correlated positively with more anxiety both at admittance (r = 0.434; p = 0.44) and discharge (r = 0.51; p = 0.17). When it comes to the 6MWT, the results improve in all pts with complete data (19 pts, average 394.14 SD 111,45 to 495,00 SD 102,89). A negative correlation was found between the initial 6MWT and depression (r = -0.46; p = 0.04). Also the Ejection Fraction correlated negatively with anxiety (r = -0.51; p = 0.02) and depression at admittance (r = -0.47; p = 0.04). At discharge an association was found between EuroQol and the 6MWT (r = 0,53; p < 0,05)
Conclusions: Further studies are necessary to establish cardiac rehabilitation programs which are aimed at these patients and to explore the variables connected to their psychosocial wellbeing.

P2555 - CARDIAC SURGERY FOR GROWN UP CONGENITAL HEART DISEASE A 12 YEAR RETROSPECTIVE FROM A TRANSITIONAL COUNTRY

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The remarkable advancements in both paediatric and adult cardiology and cardiac surgery over the past decades have resulted in an increased number of grown-ups with congenital heart defects (GUCH). This set of patients pose unique challenges and requires specialist care particularly in a transitional country. Surgical records can indicate the need for a comprehensive GUCH programme.
Study Design: We conducted a retrospective review of GUCH patients who have undergone surgery at the Groote Schuur Hospital's Chris Barnard division of cardiothoracic surgery between 1 January 2003 and 30 June 2015. We reviewed at the surgeries performed, the most common congenital heart lesion as well as the mortality rate.
Results: There were 109 cases identified as having surgery for GUCH in the 10 year study period. 61% of surgeries were performed on females and 39% on males. The largest proportion of patients, 36 patients (33%), were between 20-29 years old and the smallest proportion, 5 patients (5%), were between 50-59 years old. The majority of patients, 100 patients (92%) had corrective surgery, and 9 patients (8%) had palliative surgery.Mortality rate was 4%. The median age at death was 49. More than half, 63 (58%) of the surgeries were performed by congenital surgeons, 80 patients (73%), live in the Western Cape. The most common lesions were septal defects, seen in VSD (9%), ASD (49%), AVSD (15%), and Tetralogy of Fallot (11%). Of interest, despite the established cardiac surgery programme in the province, 92 patients (87%), had their first surgery in adulthood.
Conclusion: In conclusion, this study has shown that many people in a developing country only present for the first time in adulthood, especially with non-critical CHD lesions. The mortality rate in adulthood is low but a comprehensive GUCH service in our province with specialist teams, is needed to focus on this patient population.

P2561 - FACTORS EFFECTING QUALITY OF LIFE IN ADULTS WITH CONGENITAL HEART DISEASE IN JAPAN

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Background: International research has reported that lower quality of life (QOL) in adults with congenital heart disease (CHD) is

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associated with older age, lack of employment, never being married, and low New York Heart Association (NYHA) functional classification. However, there has been a lack of research in Japan. [Objectives] We aimed to determine the factors that affect QOL in adults with CHD in Japan.
Methods: CHD patients aged ≧ 18 years were included. We mailed self-report questionnaires that assessed factors including QOL (assessed on a linear analog scale), previous medical diagnoses, medical treatments, subjective symptoms, employment status, marital status, and age, et al.
Results: The sample consisted of 253 respondents (Response rate was 50.6%). 115 (45.5%) of which were male. Mean age was 36.4 years old. (SD 15.1). Mean QOL was 70.73 (±10.73 (1/2 SD) = 60.00, 81.46). Also we classified subjects into two groups based on QOL score (QOL < 60: low group, QOL >82: high group), and compare them on outcome variables. Low group included 69 cases, High group included 82 cases. The two groups did not significantly differ on the following factors: numbers of previous operations, age, sex, or past psychiatric disorders ($P > 0.05$). However, the low group had a higher number of hospital admissions ($P < 0.05$) and a significantly worse NYHA functional class ($P < 0.01$) than the high group. Further, fewer low group patients were married and a greater number were unemployed than high group participants ($P < 0.01$ for both).
Conclusion: The QOL of Japanese patients also associated with lack of employment, never being married and low NYHA functional class as international study, however, their age didn't relate with them. The QOL of patients having a higher number of hospital admissions were worse. We need to consider effects of their admissions and support.

P2587 - THE EFFECTIVENESS OF THE ECHOCARDIOGRAPHIC INDICES AS A METHOD OF ASSESSING THE CARDIAC FUNCTION IN ADULT PATIENTS WITH FONTAN CIRCULATION

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Background: Utility of quantitative echocardiographic assessment of left ventricular function in patients with normal heart structure have been recognized for the decision making. However, there is no useful quantitative echocardiographic index for the assessment of systemic ventricular (SV) function in patients with Fontan circulation. Therefore, SV function has been evaluated by visual grading in the clinical setting. In this study, we investigate to find out reliable echocardiographic indices which are well related with visually assessed SV function in Fontan patients.
Methods: Thirty Fontan patients (mean age 26 ± 7 years, 14 males, right ventricle (RV) in 15, left ventricle (LV) in 15) whose recorded echocardiographic images were acceptable to analyze were included and SV function were retrospectively analyzed. Following quantitative indices were analyzed: Fractional area change (FAC), Tricuspid (mitral) annular plane systolic excursion (TAPSE or MAPSE), tricuspid valve S' or mitral valve S', Global Longitudinal Strain (GLS), Global Circumferential Strain (GCS). Visual assessment of SV function was performed by two expert doctors and classified into two groups, good or reduced. Measured values were compared between visual and quantitative assessments. Receiver operating characteristic (ROC) analysis was also conducted to assess the diagnostic value of echocardiographic indices.
Results: Quantitative indices were not statistically different between visually classified groups in the single LV patients, but GCS was relatively low in visually reduced group (table). In the single RV patients, GCS was statistically lower in visually reduced group. The optimal cutoff value for GCS in ROC curve was -22% (sensitivity 89%, specificity 100%).
Conclusion: We find GCS was correlated with visually assessed SV function, particularly in single RV patients. We unconsciously assessed severity of SV function by the circumferential contraction rather than the longitude contraction in Fontan patients.

Table.

	the good function group	the reduced function group	p value
Left dominant ventricle N = 15			
MAPSE, mm	11.5 ± 1.8	9.7 ± 2.4	NS
MVS', cm/s	6.1 ± 1.0	5.7 ± 1.6	NS
FAC,%	0.46 ± 0.13	0.38 ± 0.08	NS
GLS, %	-19.8 ± 4.5	-18.0 ± 4.1	NS
GCS, %	-18.4 ± 9.5	-13.7 ± 3.5	NS
Right dominant ventricle N = 15			
TAPSE, mm	9.2 ± 3.7	8.3 ± 1.7	NS
TVS', cm/s	4.5 ± 1.4	5.0 ± 1.7	NS
FAC,%	0.40 ± 0.06	0.29 ± 0.11	0.066
GLS, %	-19.1 ± 3.4	-18.7 ± 2.7	NS
GCS, %	-23.5 ± 6.1	-11.9 ± 4.3	0.0057

P2588 - SCIMITAR SYNDROME REVISITED

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Background: Scimitar syndrome is a rare congenital disorder in which there is anomalous pulmonary venous drainage and lung hypoplasia occurring almost exclusively on the right side. In patients with a small left to right shunt management is symptomatic. In more severe cases management is surgical via re-implantation of the scimitar vein into the left atrium or insertion of an intra-atrial baffle. This syndrome encompasses a wide spectrum of presentations sometimes influenced by the presence of other congenital cardiac co-morbidities.
Methods: This case series explores the progress of three patients with Scimitar Syndrome within a single Adult Congenital Heart Disease centre over the course of seven years using data obtained from clinic letters and imaging servers. They include one male patient, age 71, and two female patients aged 26 and 40 all with right sided Scimitar veins. Their co-morbidities, presentations and treatment outcomes were outlined along with experiential commentary derived from clinic letters and the physicians involved in their care.
Results: Whilst breathlessness and initial respiratory complaints were a common theme, timing of diagnosis differed greatly. Two patients were diagnosed in infancy, the other aged 64.

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One infant diagnosis was lost to follow up for twelve years between paediatric and adult services. The late diagnosis differed in the presence of a small patent foramen ovale and required treatment with surgical intervention. The younger medically managed patients both had reassuring CPEX and 24 hour tape results, but differed greatly in their experience of symptoms and treatment plan.

Conclusions: This case series provides good examples of the broad range of presentations and outcomes in Scimitar Syndrome. It also highlights differences in patient approach to their condition; lack of correlation between clinical severity and experience of symptoms; and the importance of acknowledging this along with the challenges of continuity of care between

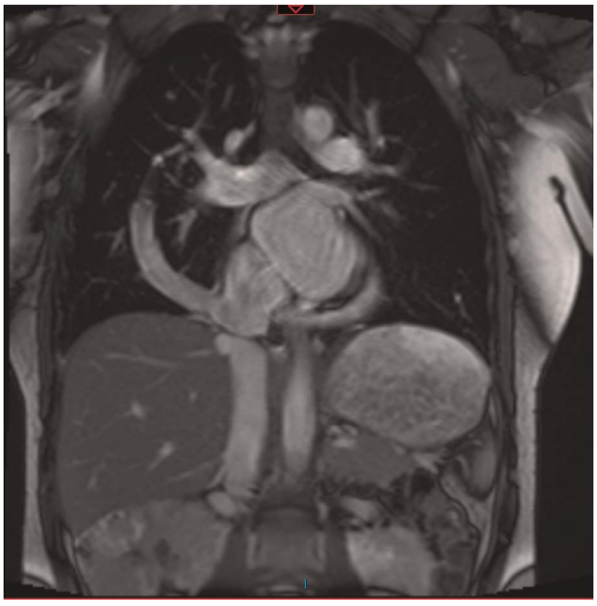

Figure 1.

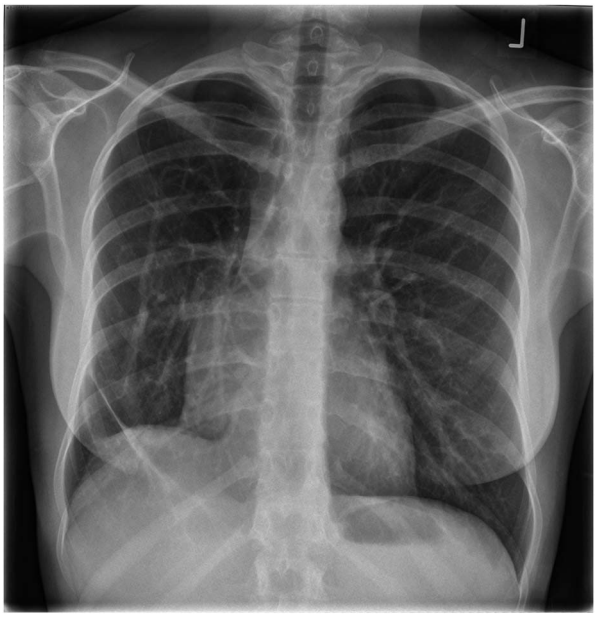

Figure 2.

P2593 - RECOARCTATION OF AORTA AFTER NORWOOD PROCEDURE IN CHILDREN WITH HYPOPLASTIC LEFT HEART SYNDROME INCIDENCE RISK FACTORS MIDTERM OUTCOME

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Background: Recoarctation of the aorta is complication after the Norwood procedure. Undiagnosed and untreated, it may lead to failure of the systemic ventricle and death. The main objective of this study is to analyze the incidence of recoarctation in our center, define risk factors and determine the outcome of patient after balloon angioplasty.

Materials and Methods: We retrospectively reviewed the records of 96 successive patients with hypoplastic left heart syndrome that underwent the Norwood (NP) procedure between 2008 and 2010. Pre-, intra, postoperative, echocardiography and angiography data were analyzed.

Results: Significant recoarctation was noted in 23 patients (32.3%) on the average of 95.1 days (49-156 days) after the NP. The ejection fraction of the right ventricle on echocardiography was decreased in the patients with coarctation as compared to the patients without recoarctation the mean decrease was 9% ($p < 0.05$). All the patients underwent percutaneous balloon angioplasty, which led to a decrease in the mean gradient at the site of narrowing from the average of 27.5 mmHg to 9.7 mmHg ($p = 0.008$) and enlarged the neoisthmus by the average of 2mm (0.7–4.5 mm) ($p < 0.05$). None of the patients needed a surgical procedure. The diameter of the ascending aorta - OR = 7.82 (95% CI [2.45-29.2]) ($p = 0.001$), atresia of the mitral valve - OR = 7.00 (95% CI [1.76-32.1]) ($p = 0.003$) and atresia of the aortic valve - OR = 6.22 (95% CI [1.69-29.4]) ($p = 0,002$) showed the highest risk of recoarctation. In the recoarctation group, the 6-year follow up was 88%, and we observed only one death before Fontan procedure.

Conclusion: An early diagnosis of recoarctation prevents the occurrence of life-threatening complications between the first and the second stage of HLHS. Balloon angioplasty is an effective intervention for treatment. In our observations, some risks factors seem to have an impact on recoarctation. Recoarctation does not increase the interstage mortality.

P2610 - PRIMARY CARE OF HEART FAILURE ARRHYTHMIAS PULMONARY HYPERTENSION AND COMORBIDITIES IN ADULTS WITH CONGENITAL HEART DEFECTS

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Background: Due to medical advances, the number of adults with congenital heart defects (ACHD) in Germany has approached 280,000. Many have chronic cardio-vascular disorders with negative effects on morbidity and mortality, including particularly heart failure, arrhythmias, pulmonary hypertension (PAH) and impeding comorbidities. Nevertheless, clinical experience shows, that in Germany more than 200.000 ACHD are not treated by

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certified specialists. The focus of this study is to demonstrate "real life" primary care of ACHD and their emerging cardiac and non-cardiac comorbidities in Germany.
Methods and Results: A questionnaire-based survey of 116 ACHD (48.3% female, mean age 34.8 ± 12.5 years) and 126 general practitioners (GP) (35.7% female, mean age 54.6 ± 9.1 years) was analysed. GP's deal with all types and severity grades of CHD, in 58% even with moderate and severe forms. ACHD consult their GPs not only for non-cardiac problems or comorbidities. Even in cardiac-related problems, more then 70% consult their GP first. GPs report to manage arrhythmias in 47%, heart failure in 48% and PAH in 22% of their consulted ACHD. More than 30% of the GPs indicate to miss sufficient support concerning specific ACHD management, e.g. regarding heart failure, PAH or anticoagulation. Frighteningly, 63.5% of the GP's are uninformed about the existence of certified ACHD-facilities, physicians and also rarely about ACHD-support groups. Although suffering from the mentioned serious problems in 41.1% ACHD are also uninformed about specific medical facilities, and ACHD-support groups (44%).
Conclusions: GPs and ACHD lack of specific disease related knowledge and skills. Both are insufficiently informed about related medical problems, about certified ACHD-facilities and patient-support groups. Better awareness and a stronger cooperation between certificated CHD-specialists and GPs as well as a better education of patients is therefore a future goal to provide adequate medical supply in order to reduce morbidity and mortality of ACHD.

P2623 - THE GROUP PROGRAM FOR PATIENTS OF TRANSITIONAL AGE

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Many textbooks and papers state that the education for patients is one of the most important issues for transition. In 2011 AHA published the scientific statement about transition and we got some guidelines for educating them, however, it is rather difficult to practice all of them. From 2010 we hold the group programs to educate young patients. The purpose of these programs is giving them the chance to talk about themselves to others and to feel and understand their own body through the experience-based learning. We plan two or three programs every year. Each program has each topic, such as sex activity and pregnancy, sports for young patients with heart disease, how to behave at school and making 'my own chart'. We select attendees taking the topic, patients' age and the type and severity of their disease into accounts. In this program, we try to shorten the time of lectures and let them talk, do exercise, play cards about their medications, experience examinations as a technician, fill their own chart by themselves or do role-playing. Over 95% of attendants feel that such programs help their understanding about their disease. Some parents say that these programs give them the chance to exchange opinions with their children about their lives and future. Some patients continue to fill a new page of their own charts at out-patients' clinic. On the other hand, it is revealed that these programs have some difficulties. Some patients whom we especially hope to attend the program are not interested in such a program. Some patients feel that it is too early to learn about sex activity and pregnancy. However we are sure that continuing the group program will help young patients' understanding about their disease and lead them to go up the stairs of transition.

P2626 - THE FATE OF RESIDUAL SHUNT IN TRANSCATHETER CLOSURE OF MULTIPLE ATRIAL SEPTAL DEFECTS

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Background: Clinical outcomes and fate of residual defects after device closure of multiple ASDs in various defect anatomies has not been clearly documented.
Methods: Patients with transcatheter closure of multiple ASDs from 2005 to 2015 were reviewed (224 patients; 17.2% of total 1295 procedures). Subjects were divided into 4 groups; Group I (n = 118, 52.6%): two nearby defects, Group II (n = 52, 22.3%): two discrete/distant defects, Group III (n = 45, 20%): multiple (≥ 3) defects, Group IV (n = 9, 4%): multi-fenestrated defects. Patients' and procedural parameters, outcomes and closure rate during follow-up were investigated.
Results: Patients' median age was 22.9 years and mean follow-up (FU) duration was 62 ± 14 months. Procedural success rate was 99.5%, 2 major complication (0.9%; 1 device embolization, 1 complete heart block) and 4 minor complications (1.8%) occurred. Overall, the residual shunts tended to reduce in size or close spontaneously with time. Complete closure rates after procedure and at last FU in each group were as follows; group I – 84,7% and 98%, group II – 9.6% and 90%, group III – 11.1% and 92%, group IV – 10.3% and 87.5%. A hundred sixteen (98%) in Group I and 44 (84.6%) in Group II patients were treated using a single device. In these groups, complete closure was related with use of oversized device. In Group III, mean 1,4 devices were used. (2 devices in 12 patients, 3 devices in 3 patients) All the residual shunts were small (≤3 mm) without hemodynamic significance.
Conclusion: Transcatheter closure is an effective and safe therapeutic option in patient with multiple ASDs. Small residual shunt tends to reduce in size or closed spontaneously according to the FU duration. Use of a slightly oversized device may facilitate complete closure rate in patients especially with 2 defects treated by a single device.

P2629 - BODY MASS INDEX AND CO MORBIDITIES IN A COHORT OF ADULT FONTAN PATIENTS

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Yale University School of Medicine, Pediatrics/Internal Medicine, New Haven-United States[1]; Yale University School of Medicine, Internal Medicine, New Haven-United States[2]; Yale University School of Medicine, Pediatrics, New Haven-United States[3]

Background: The Fontan procedure is palliative for patients with functionally univentricular hearts. Though obesity is prevalent among children with congenital heart disease (CHD), children post-Fontan are thought to have lower rates of obesity compared to other CHD groups. Whether that remains true in adults is unknown.
Methods: A retrospective review was conducted of adult Fontan patients seen at Yale-New Haven Adult Congenital Heart Program since 2013. Clinic records were reviewed for surgical and medical history. Body mass index (BMI) was calculated from height and weight at most recent visit. Overweight/obese patients (BMI >25) were compared to non-obese group.
Results: 44 patients (29.5 yrs, 18-55) were reviewed, 20 (45%) of which were female. Mean time since Fontan operation was 23.6

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yrs (5-37). There were 24 lateral tunnel, 10 atriopulmonary and 10 external conduit Fontan operations. Using National Heart, Lung and Blood Institute classification system for BMI, 6 patients (13.6%) were underweight (<18.5), 13 (29.5%) were normal weight (18.6-24.9), 14 (31.8%) were overweight (25.0-29.9), 6 (13.6%) had Class I Obesity (30.0-34.9), 3 (7%) had Class II Obesity (35.0-39.9) and 2 (5%) had Class III extreme obesity (>40.0). The BMI range was 16.5-58.9, median was 26.3. Overweight/obese Fontan patients used more anti-hypertensive medications than non-obese group, 1.04 vs. 0.63, $p = 0.019$. One patient developed Type 2 diabetes and one underwent sleeve gastrectemy for morbid obesity. For the sub-set with recent catheterization ($n = 23$), there were no significant differences for mean wedge or Fontan pressure between the two groups.
Conclusions: In a cohort of adult Fontan patients, less than 1/3 are in a healthy BMI range. 54% were overweight or obese. Use of anti-hypertensive medications was higher in obese Fontan patients. Given the risk for development of co-morbidities, adult Fontan patients may benefit from closer monitoring for complications along with targeted weight loss interventions.

P2646 - VENTRICULAR RESPONSE TO DOBUTAMINE STRESS IS A PREDICTOR FOR OUTCOME IN PATIENTS WITH REPAIRED TETRALOGY OF FALLOT

Eva Van Den Bosch[1], Judith Cuypers[2], Saskia Luijnenburg[1], Jochem Van Den Berg[1], Nienke Duppen[1], Eric Boersma[2], Jolien Roos-hesselink[2], Willem Helbing[1]
Erasmus MC, Sophia Children's Hospital, Department of Pediatric Cardiology, Rotterdam-The Netherlands[1]; Erasmus MC, Department of Cardiology, Rotterdam-The Netherlands[2]

Background: Outcome after the surgical correction of tetralogy of Fallot (ToF) is good. However, severe pulmonary regurgitation (PR) and progressive right ventricular (RV) dilatation and dysfunction are frequent causes of late morbidity and even mortality. Optimal timing of pulmonary valve replacement (PVR) for severe PR remains difficult. Evaluation of left and right ventricular function using exercise or pharmacological stress cardiac magnetic resonance imaging (CMR) has been advocated as a tool to reveal impending cardiac dysfunction. Several studies have shown that patients with ToF have abnormal RV stress response. Our aim was to evaluate the prognostic value of dobutamine stress CMR in ToF patients.
Methods: Between 2002 and 2012 Fallot patients prospectively received low dose dobutamine (7.5 µg/kg/min) stress CMR. Cardiac events (reintervention, arrhythmias or cardiac death) during follow-up were recorded. Time-to-event analyses (Kaplan-Meier method) and Cox proportional hazard regression analysis was performed to determine the predictive value of dobutamine stress CMR.
Results: 86 patients (55 male, median age 19.5 (7.4-49.3) years) underwent stress CMR. Overall indexed left and right diastolic volume decreased during stress (RVEDVi from 130.5 ± 41.7 ml/m^2 (rest) to 125.6 ±14.3 (stress) and LVEDVi from 79.3 ±12.9 (rest) to 76.6 ±40.2 (stress)) and right (RVEF) and left (LVEF) ejection fraction increased. After a median follow-up of 11.1 years 33 patients (38%) developed an event The event-group showed a significantly lower increase in RVEF fraction during stress (right ventricular reserve) compared to the non-event-group ($9.2 \pm 3.8\%$ vs $12.6 \pm 6.1\%$, $p = 0.002$). Multivariate Cox regression analysis showed right ventricular reserve (HR 0.88; 95% CI 0.82-0.94 ; $p < 0.001$) and RVEF at rest (HR 0.90; 95% CI 0.85-0.95; $P < 0.001$) to be associated with risk of an event, corrected for age.
Conclusion: Right ventricular response to dobutamine stress seems to be an additional predictor for events in patients with repaired ToF.

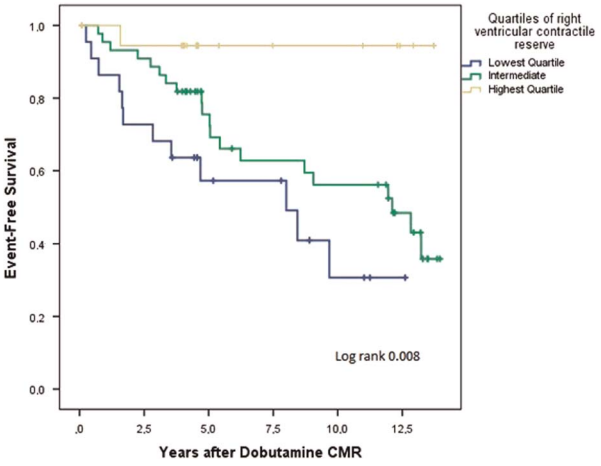

Figure 1.
Kaplan-Meier curve for event-free survival.

P2660 - PER CUTANEOUS PULMONARY VALVE IMPLANTATION OUR EXPERIENCE

Ramya Shri Chandrasekaran, Amol Gupta, Sivakumar Kothandam
MMM, Pediatric Cardiology, Chennai-India

Background: Corrective surgery for heart diseases with valve conduits needs redo surgeries for replacement of failing conduits. Percutaneous pulmonary valve implantation (PPVI) into these failing conduit is less morbid alternative to surgery.
Method: Patient's demography,symptoms,echo,cardiac MRI and other imaging modalities were analyzed.PR defined by color Doppler and MRI, RV dysfunction and dilatation assessed by echo and MRI.Balloon sizing of the RVOT and simultaneous coronary angiography performed.PPVI done using 22mm straight and 34mm flare designed self-expanding porcine transcatheter valve. After deployment valve function is assessed by echo, hemodynamic data,and angiography.On follow up patients reassessed with echo at regular intervals within first year and cardiac MRI at 6 months and 1 year to assess the competency of valve.
Results: PPVI was performed successfully in 6 patients with a median age of 24.1(12-37) years and median weight of 46.25 (41.6-77) kg.The majority had Tof with pulmonary atresia (n 5), post Ross procedure (n 1).conduit was bovine pericardial valved conduit in 1,aortic homograft in 3, PTFE conduit with pericardial monocusp in one.1 patient had native RVOT repaired by transannular patch.Interval between surgery and PPVI was 5-17 years, RVOT stenting performed before PPVI with a stainless steel (n 3) & platinum iridium stents (n 2) .Except 1 patient with 34mm flare design venus P valve, rest of the patients had 22mm straight cell venus P valve was implanted. No procedural complications Follow up was 7 to 810 days with median of 180 days.Cardiac MRI was performed Post PPVI in 2 patients, in others MRI was contraindicated due to previous RVOT stenting. It showed fall in RVEDVi (from 135.5 to 94.56.ml/m2) and PR RF (from 28.516.2 to 53.53).
Conclusions: PPVI resulted in the ability to avoided multiple surgical right ventricular outflow tract revisions.

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P2665 - A WOMAN OF POLYSPLENIA WHO RECOVERED FROM CYANOSIS CAUSED BY PULMONARY ARTERIOVENOUS MALFORMATIONS WITH THE RECONSTRUCTION IN 15 YEARS AFTER THE FIRST TCPC OPERATION

Minako Hoshiai[1], Hiroaki Kise[1], Takako Toda[1], Keiichi Koixumi[1], Shigeru Tateno[2], Kozo Matsuo[3]
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Background: Pulmonary arteriovenous malformation (PAVM) is one of the major complications in the patients operated with the staged approach to total cavoplumonary connection (TCPC). We report a polysplenia syndrome woman who required surgery intervention again because of the cyanosis progressed rapidly, 15 years after the first TCPC.
Case: She was 18 years old. The diagnoses of her heart anomaly complex were single ventricle, pulmonary stenosis, left side superior vena cava, interrupted of the inferior vena cava with hemiazygos vein continuity. She had a TCPS (LSVC-LPA connection) operation at 16 months old and had TCPC which was hepatic vein and right pulmonary anastomosis at 3 months for 2 years old. The desaturation in her was aggravated rapidly at 16 years old, and her status became to NYHA III because of PAVM in left lung in 15 years after the first TCPC. We considered the effective way for hepatic vein flow influx into LPA. Then, we connected the conduit from hepatic vein to right side innominate vein, for mixing of all venous return. The 3-month follow-up data supported this choice with a clear improvement in the overall clinical condition and oxygen saturation levels that had increased from 72% to 91%, implying regression of the PAVMs.
Discussion and Conclusion: The pathophysiology of PAVM is lack of hepatic factor in pulmonary artery because without hepatic direct perfusion in TCPC. This case had passed from the first TCPC for a long time, but the cyanosis improved in a short period from this operation. Praus et al reported that the only hepatic venous blood closed PAVMs after cavopulmonary connection, Even the case which has passed a long period from the first TCPC operation in the first time like this case considers a re-operation to PAVM aggressively.

P2669 - CARDIAC REMODELING AFTER PERCUTANEOUS CLOSURE OF ATRIAL SEPTAL DEFECT IN ADULTS AN ECHOCARDIOGRAPHIC STUDY WITH NEW TECHNIQUES

Danielle Rocha[1], Carlos Augusto Pedra[1], Simone Pedra[1], Jorge Assef[2], Valmir Fontes[1]
Dante Pazzanese Institute of Cardiology, Pediatric Cardiology, Vitoria-Brazil[1]; Dante Pazzanese Institute of Cardiology, Ecocardiography, Sao Paulo-Brazil[2]

Introduction: The atrial septal defect of the secundum type (ASD)is a frequent congenital heart disease,being the most commonly encountered in the adult population. New echocardiographic techniques have been employed to assess the dimensions, geometry and function of the cardiac chambers. Objectives: Hypothesizing that percutaneous closure,results in a fast cardiac remodeling even in adults with chronic volume overload of the right chambers. Our aim was to assess the temporal pattern of cardiac remodeling and analyse possible differences between patients of different age groups and different ASD sizes.
Material and Methods: This was a study of a cohort of twenty nine adults submitted to percutaneous closure of the ASD with the Cera device followed along a year. Cardiac remodeling was assessed by various echocardiographic techniques. Bidimensional echocardiography was used to measure the dimensions of the right atrium (RA), right ventricle (RV) and left ventricle (LV), to determine the fractional area of the RV (FAC), and to analyse the volumes of the cardiac chambers. Three-D echo was employed for volumetric and functional analysis of the RV. Acoustic speckle tracking was utilized to assess the function of the RA, RV and LV.
Results: The ASD dimension and the size of the device was a mean of $20{,}2 \pm 5{,}0$ mm and $22{,}9 \pm 6{,}2$ mm, respectively. After the procedure, there was an early reduction of the RA (<0.001) and RV (<0.001) sizes and an increase of the LV dimensions ($p < 0.014$). There were no significant changes in the function of the RA as assessed by longitudinal strain ($p = 0.227$ for the P wave and $p = 0.124$ for the T wave). There was an abrupt reduction of the RV function assessed by longitudinal strain ($p = 0.002$) and 3D echo ($p = 0.084$). There was an immediate decrease in the RV stroke volume ($p < 0.001$) and an increase in the LV stroke volume ($p = 0.027$). There was an immediate increase in the LV function as assessed by the Simpson method ($p = 0.052$) and a reduction in its longitudinal strain ($p = 0.049$).
Conclusions: Percutaneous closure of the ASD in middle aged adults results in fast cardiac remodeling sustained results over the first year of follow up.

P2678 - SURGICAL REPAIR OF VENTRICULAR SEPTAL DEFECT IN ADULTS OUTCOMES AND INDICATION

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Objectives: Outcome data of VSD (ventricular septal defect) closure in adults on which to base recommendations are limited. We sought to review our experience with surgical VSD closure in adults
Methods: We retrospective reviewed 126 patients (mean age 33.7 ± 12.5, 77 male) who underwent surgical VSD closure (1996 to 2016)
Results: VSDs were classified according to the Society of Thoracic Surgeons as type 2 (n = 58, 46.0%), type 1 (n = 49, 38.9%), and type 4 (n = 19, 15.1%). Aortic cusp prolapse (n = 49, 38.9%), aortic regurgitation (AR) (n = 33, 26.2%), Qp/Qs ratio ≥ 2 (n = 22, 17.5%), pulmonary hypertension (n = 15, 11.9%), left ventricular volume overload (n = 14, 11.1%), sinus of Valsalva rupture (n = 11, 8.7%), and infective endocarditis (n = 9, 7.1%) were indications for closure. Aortic valve repair and aortic valve replacement were performed in 20 patients and 5 patients, at the same time of VSD closure. Mean follow-up duration was 4.6 ± 4.2 years. One patient, who underwent VSD fenestration after VSD closure due to severe pulmonary hypertension, died postoperatively cause of pneumonia and septic shock. Reoperation included aortic valve replacement in 2 patients, repair of residual defect in 2 patients, and VSD fenestration in 1 patient, described as above. At 10 years, freedom from of reoperation was $91.6 \pm 4.3\%$. Late moderate AR developed in 7 patients at mean 5.5 ± 2.6 years (5 of 7 received aortic valve repair at VSD repair). Preoperative greater than moderate AR was risk factor for late moderate AR in univariate analysis ($p = 0.002$).
Conclusions: VSD closure in adults was performed with low mortality and morbidity. We consider that VSD with aortic valve deformity or AR should be treated aggressively before progressing.

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P2716 - PULMONARY VEIN STENOSIS A SYNOPSIS OF A CASE MANAGED IN COMPARISON TO CONTEMPORARY LITERATURE

Venkatram Subramanian, Arun Karunakaran, Ashish Shah, Petra Jenkins, Jaspal Dua, Heiko Schneider
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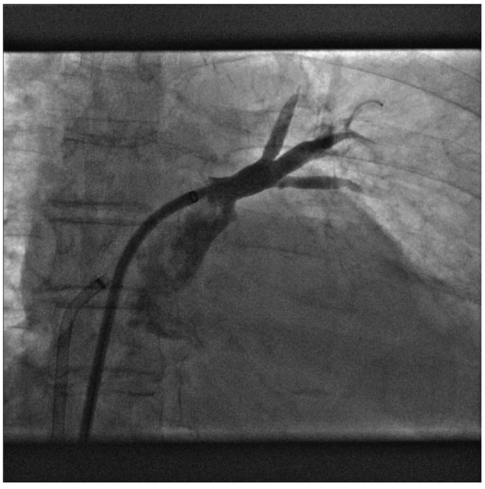
Background: This report describes the successful treatment of pulmonary vein stenosis which arose as a complication successful catheter ablation of atrial fibrillation.
Methods and Results: Clinical and operative notes of a patient managed for Pulmonary Vein stenosis in Manchester Royal infirmary with comparison of similar cases and review of literature regarding the management of Pulmonary vein stenosis. Pulmonary vein stenosis following ablation of atrial fibrillation is a known complication. Development of pulmonary vein stenosis resulted in significant rise in pulmonary artery pressures and pulmonary hypertension. Patient had dyspnoea on exertion limiting daily activities significantly. Echo cardiogram followed by CMR and right and left heart catheter was performed confirming the diagnosis. The Left pulmonary vein showed severe stenosis and therefore offered stenting of pulmonary vein. Although balloon angioplasty only resulted in mild improvement in pulmonary artery pressures subsequent pulmonary vein stenting resulted in near normal pulmonary artery pressures and marked symptomatic improvement in dyspnoea.

Conclusion: Pulmonary vein stenosis is an uncommon but potentially life-threatening complication of ablation of the pulmonary veins. There is a discrepancy in the definitive method of tackling the stenotic vessel especially in the adult population. Overall consensus on early intervention in stenosis >75% has been advised. Symptomatic benefit may be obtained by balloon dilation and in selected cases, stenting of the pulmonary vein. Although the long-term outcomes are unknown, in the absence of other effective modalities, pulmonary vein stenting should be considered.



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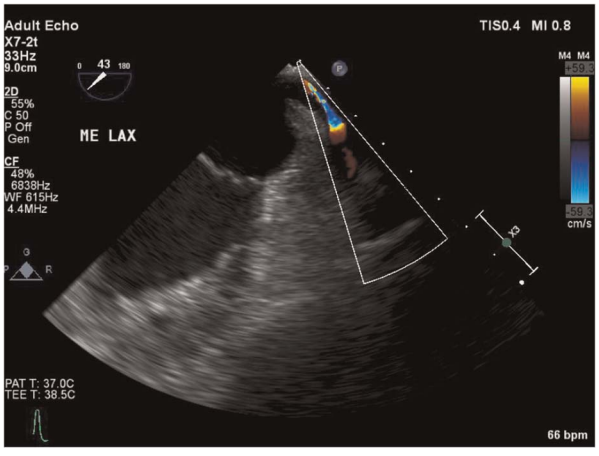

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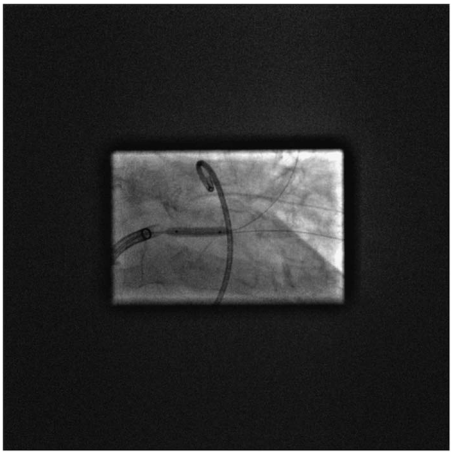

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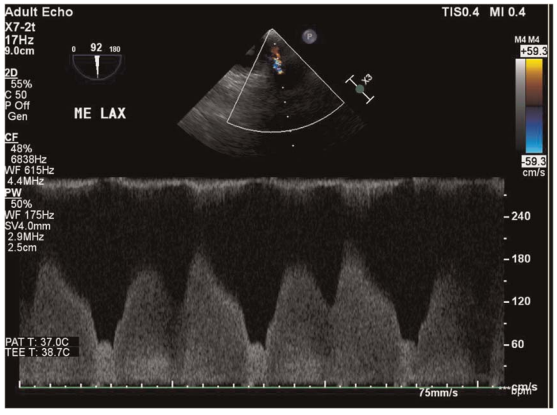

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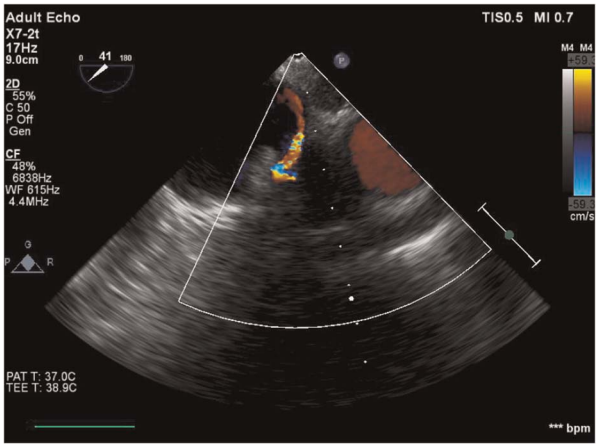

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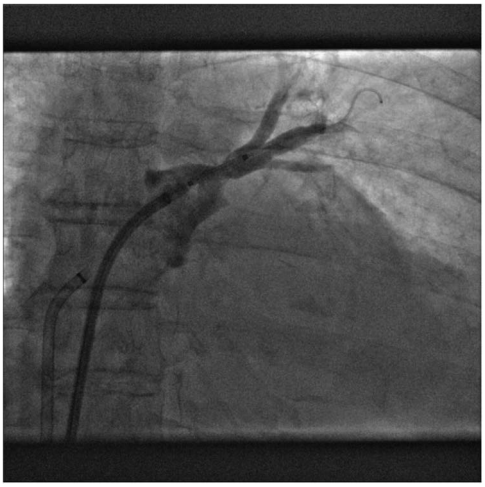

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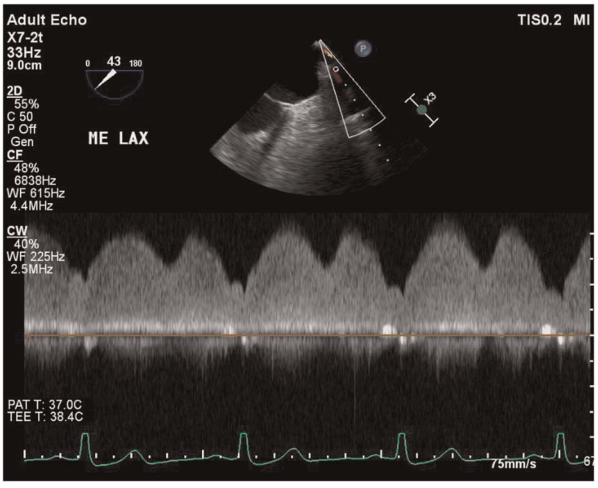

Figure 7.

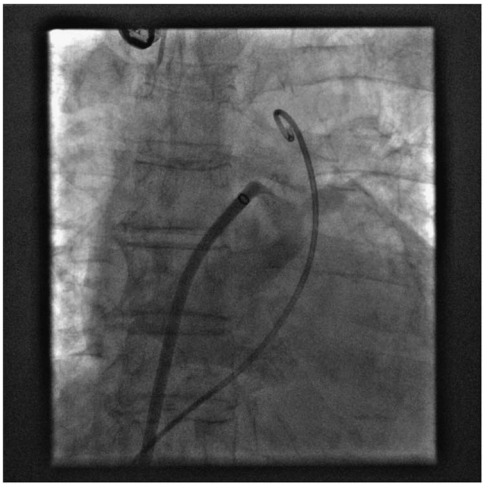

Figure 8.

P2728 - CASE SERIES A RARE CASE OF ADULT CONGENITAL HEART DISEASE WITH DEXTROCARDIA UNDERGOING DEFINITIVE OPERATION IN FIVE YEAR PERIOD IN DR. SOETOMO GENERAL HOSPITAL SURABAYA (JANUARY 2012 DECEMBER 2016)

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Introduction: Dextrocardia is mostly associated with congenital cardiac defect that is the heart positioned at right thoracic cavity. Dextrocardia itself is a rare congenital cardiac defect with prevalence of 1:12,000. In general cardiac center,adult congenital heart disease with dextrocardia is also a rare case in congenital heart disease population approximately 1.6%. Cardiac surgical procedure done in adult congenital heart disease with dextrocardia has many challenges, not only the position of surgeon, but also the existing complication regarding adult age.
Objectives: We aimed to report serial case of adult congenital heart disease with dextrocardia managed with definitive operation in our center for 5-year period, since January 2012 until December 2016.
Result: We had 12 patients with dextrocardia (1.9%) among 612 patients with congenital heart disease undergoing cardiac surgical procedure in our center for 5-year period. There were 4 patients (0.6%) adult congenital heart disease with dextrocardia treated surgically in our center for 5-year period. We had done definitive surgeries for those patients, those are 2 patients Tetralogy of Fallot undergoing total correction, 1 patient large subaortic ventricular septal defect (VSD) with bidirectional shunt and pulmonary hypertension undergoing VSD closure by double flap patch and 1 patient Lutembacher Syndrome with pulmonary hypertension undergoing mitral valve replacement and atrial septal defect closure. Those four patients were successfully treated surgically. The eight remaining patient dextrocardia underwent palliative procedures.
Conclusion: The challenges of adult congenital heart disease with dextrocardia are position of surgeon during operation in first assistant (left position of patient) complicating surgeon's orientation in repairing the defect and postoperative complication regarding the physiologically changed hemodynamic status due to long-standing congenital malformation. It requires special attention in postoperative preparation including hemodynamic status of patients and surgical,technique regarding orientation, of defect.

P2738 - EBSTEIN'S ANOMALY OF THE TRICUSPID VALVE 13 YEARS OF EXPERIENCE IN A TERTIARY CENTRE FROM FETUS TO ADULT

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Kingdom[6]; Guy's And St Thomas' NHS Foundation Trust; King's College London, Adult Congenital Heart Disease, London-United Kingdom[7]

Background: Ebstein's anomaly (EA) is a rare congenital heart defect. Patients often present with malfunctioning TV and right ventricular (RV) dilatation, which may require intervention. The timing of intervention and its outcome are still uncertain.
Method: All patients with EA followed-up in our centre between 01/01/2004-25/12/2016 were identified from the Departmental databases. Demographic characteristics, NYHA class, cardiac MRI, echocardiography, and CPET at their last follow-up were reviewed. For patients who underwent TV surgery, pre- and post-procedure data were analyzed.
Results: Of the 35 patients prenatally diagnosed, 8 were terminated, 2 died before and 6 immediately after birth. Only patients with isolated EA or with an associated atrial septal defect were included (n = 90, median age 21.8 yr, range 1.2 months-81.6 yr). Eleven patients required a systemic-to-pulmonary artery shunt or Glenn/Hemifontan (median 2.58 yr, range 4 days–63.47 yr); of these 3 underwent completion of Fontan circulation. Twenty-two patients required TV repair (n = 18) or replacement (n = 4) for severe regurgitation (median age 18.8 yr; range 1.78 yr-64.25 yr). After TV repair, there was a significant reduction in TV regurgitation (TVRF from 57 ± 16% to 16 ± 13%; $p < 0.001$), the RVEDV decreased (from 176 ± 81 ml/m^2 to 114 ± 26 ml/m^2; $p = 0.06$), the RV effective stroke volume (RVSV) increased (from 32 ± 13 ml/m^2 to 48 ± 20 ml/m^2; $p = 0.19$), the LVEDV volume increased (from 50 ± 14 ml/m^2 to 76 ± 17 ml/m^2; $p = 0.03$) and the LVSV increased as well (from 31 ± 8 ml/m^2 to 43 ± 9 ml/m^2; $p = 0.04$). These results were associated with improvement in symptoms (NYHA I from 0% to 60%; $p = 0.01$) which was mirrored by improved VE/VCO2 slope (from 34.5 ± 7.4 to 26.7 ± 4.1; $p = 0.08$). There were 1 post operative and 5 late deaths.
Conclusion: EA is characterized by a very broad spectrum of disease severity. Most severe cases require univentricular palliation. Among the others those with severe TV regurgitation are likely to receive TV repair. After surgery TV regurgitation decreases and the LVEDV increases significantly reflecting a better preload; symptoms and exercise capacity improve.

P2748 - VENTRICULAR RESPONSE TO DOBUTAMINE STRESS IS A PREDICTOR FOR OUTCOME IN PATIENTS WITH A FONTAN CIRCULATION

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Background: Despite improved short-term survival the Fontan circulation has serious long-term complications including circulatory failure, thromboembolic events and arrhythmias. Evaluation of ventricular function using exercise or pharmacological stress cardiac magnetic resonance imaging (CMR) has been advocated as additional tool to reveal impending Fontan dysfunction. Several studies have shown that patients with a Fontan circulation have an abnormal response to dobutamine stress. Our aim was to evaluate the prognostic value of dobutamine stress CMR in Fontan patients.
Methods: Between 2004 and 2012 Fontan patients prospectively received low dose dobutamine (7.5 μg/kg/min) stress CMR. After the baseline examination patients were followed up, cardiac events (reintervention, arrhythmias or cardiac death) were recorded. Time-to-event analyses (Kaplan-Meier method) and Cox proportional hazard regression analysis was performed to determine the predictive value of dobutamine stress CMR.
Results: 74 patients (49 male, median age 11.3 (6.7-24.2) years, median time after Fontan completion 7.9 (4.1-21.3) years) underwent stress CMR. Overall indexed end systolic volume (ESVi) and indexed end diastolic volume (EDVi) decreased significantly during stress (ESVi 39.9 ± 16.1 (rest) to 27.1 ± 13.6 (stress), EDVi 86.4 ± 21.3 rest to 74.0 ± 21.4 (stress)). The indexed stroke volume(SVi) did not change during stress. After a median follow-up of 5.8 years 22 patients (30%) developed a cardiac event, median 12.6 (7.6-26.5) years after Fontan completion. The event-group showed a significantly lower increase in ejection fraction (EF) during stress (ventricular reserve) compared to the non-event-group (6.1 ± 7.5% vs 11.8 ± 6.3%, $p = 0.002$). Multivariate Cox regression analysis showed ventricular reserve (HR 0.87; 95% CI 0.81-0.93; $p < 0.001$) is associated with the risk of an event, corrected for age at stress CMR and EF at rest.
Conclusion: Ventricular response to dobutamine stress may be an additional predictor for events in patients with a Fontan circulation.

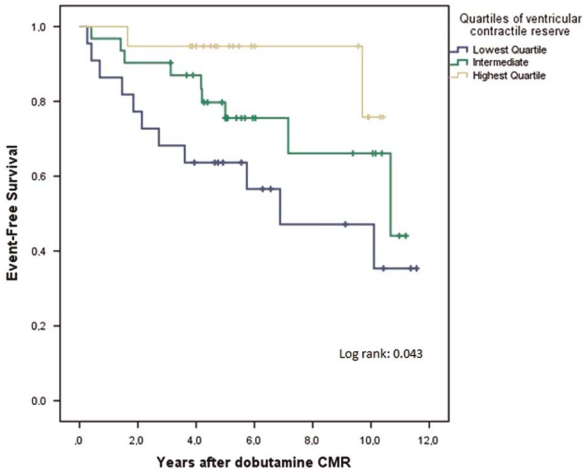

Figure 1.
Kaplan-Meier curve for event-free survival.

P2763 - ANOMALOUS CORONARY ARTERIES FROM PULMONARY ARTERY (ALCAPA & ARCAPA) SPECTRUM A SINGLE ACHD CENTRE EXPERIENCE

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Background: Anomalous origin of Left (or Right) coronary artery from pulmonary artery (ALCAPA /ARCAPA) syndrome (Bland-White-Garland Syndrome) is rare, less than 0.5% of all congenital heart defects. In 5% of cases, it is associated with cardiac anomalies e.g. atrial septal defect, ventricular septal defect, aortic coarctation. ALCAPA can result in "coronary steal" phenomenon, where left-to-right shunt causes abnormal left ventricular

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perfusion. If untreated, up to 90% with ALCAPA syndrome die within 1st year of life. Those who reach adulthood may suffer myocardial infarction (MI), left ventricular systolic dysfunction (LVSD), mitral regurgitation (MR), silent myocardial ischaemia, malignant arrhythmias, causing sudden cardiac death. Early diagnosis/prompt surgical intervention, aiming to restore two-coronary-artery circulation can lead to myocardial recovery. Most literature pertaining to ALCAPA are paediatric cases or individual case studies

Methods: We present our single centre experience of ALCAPA/ARCAPA, presenting in adulthood. Electronic search of the departmental database/retrospective case-note review was undertaken. Over a 7 year period, we had 9 ALCAPA/ARCAPA patients.

Results & Discussion: Presentations varied from syncope, to angina, to increasing dyspnoea. Two patients presented with MI, 4 with atrial/ventricular dysrhythmias, 2 with MR, 1 with severe tricuspid regurgitation, 3 with, at least moderate LVSD and 1 with pericardial effusioN. As for outcomes, 3 patients underwent coronary artery corrections; 1 had mitral valve repair; 1 had tricuspid valve repair; 1 had valve conduit repair; 1 had pulmonary patch repair and one had pericardiocentesis. 2 patients had EPS/ablation; 2 had implantable loop recorders;, 1 received an implantable cardioverter-defibrillator and 3 had permanent pacemakers. Two patients are awaiting surgery. Echocardiographic findings included LVSD, MR and lesions e.g. tetralogy of Fallot

Conclusions: ALCAPA/ARCAPA is rare. Presentation in adult age can vary. Management depends upon initial presentation/concurrent issues. Clinicians should be aware of this rare, but potentially treatable anomaly.

P2885 - LONG TERM OUTCOME OF PROTEIN LOSING ENTEROPATHY IN CARDIAC DISEASE

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Introduction: Protein-losing enteropathy (PLE) is a devastating complication of cardiac disease, especially after Fontan operation. The purpose of this study was to investigate the clinical characteristics and outcome of PLE patients at a single institution.

Methods: We reviewed medical records of 34 patients with PLE from cardiac disease from 1992 to 2016.

Results: Median age at PLE diagnosis was 11.4 years (range 0.8-28.3). The follow-up duration was 7.7 ± 5.8 years. The underlying cardiac disease was functional single ventricle in 26 patients (76%), constrictive pericarditis in 3 patients (9%), valvular heart disease in 1 patient (3%), and restrictive cardiomyopathy in 1 patient (3%). Most patients (73%) underwent Fontan operation and 5 patients (14%) did not receive any surgery. PLE occurred in 4.5% of patients after Fontan. The survival rate was 80.7% at 5 years and 73.9% at 10 years. Twelve patients died during follow-up in 6.9 ± 5.9 years after PLE onset. Aortic oxygen saturation < 90%, hemoglobin level < 12 g/dl, decreased ventricular function, NYHA functional class III or IV were predictors of mortality in PLE patients after Fontan operation. For the management of PLE, medical treatments were more frequently used including diuretics, ACEI/ARB, subcutaneous heparin injection, oral corticosteroids. Interventional and surgical therapies such as Fontan pathway fenestration creation, Fontan conversion, and Fontan takedown surgery were applied in selected patients. One third of PLE patients after Fontan operation showed resolution of PLE. In Fontan patients, resolution of PLE was achieved by heparin in 4 patients, surgical Fontan fenestration in 2 patients, Aorto-pulmonary collaterals surgical ligation and transplantation in 1 patient each. Higher Fontan pathway pressure was associated with intractable PLE.

Conclusions: The survival of PLE with cardiac disease has improved with the advancement of the conservative care. Further investigation is needed to determine the individual susceptibility of PLE as well as to develop new method of prevention and therapy.

P2907 - ATRIAL SEPTAL DEFECTS IN ADULTS. EXPERIENCE OF SURGICAL RESULTS IN CARLOS ANDRADE MARÍN HOSPITAL IN ECUADOR FOR 13 YEARS. ¿LOW OR HIGH MORBIMORTALITY

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Introduction: The population of adults with congenital heart disease has increased in recent decades. Congenital heart disease, atrial septal defect (ASD) type, can lead to symptoms such as exercise intolerance with significant disability in a previously asymptomatic patient. Surgical repair has low mortality at an early age (<1% in patients without significant comorbidity) and good long-term results.

Subjects and Methods: We conducted a retrospective study, considering all cases of atrial septal defect underwent surgery in adults (>18 years), from January 2003 to June 2015, assessing the surgical benefit in the short and medium term, and evaluating improvement, deterioration or maintenance of functional class, determination of residual shunt, size and function of the right ventricle, tricuspid regurgitation and pulmonary artery pressure by transthoracic echocardiography and assessment of arrhythmias known or background.

Results and Discussion: A total of 163 patients were collected from all cardiac surgeries performed in the period described, of which the most representative group was the women (n = 114) compared to males (n = 49). Surgical Benefit: it was 100% in all operated without mortality data; Functional Class: 14 patients returned to functional class II to I (28% improvement in functional class II), 3 patients underwent functional class III to II (100% improvement in NYHA functional class III), and 17 patients joined functional class I, from the previous 110 patients who had; Right Ventricular Function (TAPSE): improved in 17 cases after surgery, with 9 patients within the first three months and 8 patients at 6 months; whereas Systolic Pulmonary Artery Pressure (SPAP): improved in 26 cases. Documented 4 types of Arrhythmias

Conclusions: Surgical repair of an ASD in adults in our institution has a zero mortality and good results with normal life expectancy and low morbidity in the short, medium and long term.

P2913 - THE RISK OF CHRONIC DISEASE IN ADULTS WITH CONGENITAL HEART DISEASE A NATIONWIDE POPULATION BASED WITH CASE CONTROL STUDY IN TAIWAN

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Background: The relationship between congenital heart disease (CHD) and chronic disease has not been determined. The purpose of this study was to examine the prevalence of major chronic diseases in adult CHD patients and to compare it with healthy adult in Taiwan.

Materials and Methods: Adult CHD patients (> = 18 years) and healthy adults were identified from the Taiwan National Health Insurance Research database between 2010 and 2013. Outcome measures were the prevalence of 22 selected chronic diseases and the log-rank test was used to evaluate the survival curves of recorded chronic diseases.

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Results: A total of 1746 adult CHD patients and 6984 healthy controls matched by age, gender and region of residence were identified and included in current study. The overall crude prevalence of adult CHD was 0.523 to 0.918 per 1000 adults with a mean age of 37.8 years (58.7% female). Top four common chronic diseases in adult CHD patients were hypertension (24.7%), cardiac dysrhythmias (20.5%), angina and coronary heart disease (18.0%), hyperlipidemia (15.5%), and chronic obstructive pulmonary disease (15.3%). Cases were significantly more likely than controls to have all selected chronic diseases ($p < 0.05$).
Conclusions: Adults with CHD have significantly increased chronic disease risk, especially cardiac and lung disease. Health care professionals should develop long term care guideline for adult with CHD to prevent chronic disease.

P2921 - SINGLE ANOMALOUS PULMONARY VEIN CAUSING SIGNIFICANT EXERTIONAL DYSPNEA IN A 60 YEAR OLD MAN

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Anomalous drainage of single pulmonary vein rarely causes symptoms. We present 60 year-old with improvement in dyspnea on exertion after surgical repair. He is a 40 pack year ex-smoker with history of treated CAD, obesity and hypertension. Additional history of inhalational farm injury treated with a nebulizer, and oral steroids with complete resolution. Presented to an ED with complaints of acute onset of dyspnea on exertion. Chest CT revealed anomalous drainage of the left upper lobe pulmonary vein to the brachiocephalic vein, no PE, and minimal centrilobular emphysema in the upper lobes. A complete set of pulmonary function tests were normal except for minimal reduction in DLCO (75%). Echocardiogram revealed mild right heart dilatation and moderate LV diastolic dysfunction. 18 months before this episode he was diagnosed with CAD treated with 3 RCA stents. Nuclear stress test showed no evidence of cardiac ischemia. After unsuccessful trial of newer medications for COPD he had repeat cardiac catheterization. Coronary angiography was normal with widely patent coronary arteries including the previous RCA stents. The LUPV drained to the lateral innominate vein without obstruction and was quite large angiographically. Innominate vein and SVC were dilated. Qp:Qs 1.2 to 1, mean PA pressure 26-28, PCWP 14. Surgical correction was undertaken with successful reimplantation of LUPV to LA appendage and closure of PFO. The surgeon noted a very large LUPV. Post-op he had complete resolution of his debilitating exertional dyspnea. Despite the fact that a single anomalous pulmonary vein usually results in no symptoms, occasionally it can lead to symptomatic pulmonary overcirculation requiring corrective surgical intervention. Perhaps co-morbidities such as coronary artery disease, primary lung disease and/or diastolic LV dysfunction could be contributing factors in such a circumstance. Patients with clinical symptoms and single anomalous pulmonary vein should be considered for surgical correction.

P2934 - CANADIAN MUSTARD AND SENNING POPULATION DEMOGRAPHICS AND MANAGEMENT

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Background: There exists no national data on the demographics, follow-up and management of patients that have underwent a Mustard or Senning procedure in Canada. Furthermore, little is know on the growing number of patients in heart failure and pulmonary hypertension.
Methods and Results: A scenario-based, self-administered survey was completed by a congenital cardiac surgeon from each of the 8 congenital cardiac surgery programs across Canada with the collaboration of their multi-disciplinary team. Half of programs followed more than 50 Mustard patients (Range: <20-280 patients) and less than 20 patients who underwent a Senning (Range: 0-100 patients). Less than 5 Mustard and Senning patients have cardiac surgery per year. Respondents reported that a total of 28201 adult congenital cardiac patients are actively followed across Canada (mean: 3512 + /- 356 patients/program). A first scenario was: "A 35 year-old male patient who underwent a Mustard procedure is deemed clinically stable." Respondents (5/8) would evaluate the Systemic Right Ventricle (sRV) yearly: TTE (7/8), MRI (6/8), TEE (1/8). The second scenario was: Same patient however, last TTE showed a 40% sRV Ejection fraction with moderate systemic Tricuspid Regurgitation (sTR). Respondents (8/8) claimed they would evaluate sRV every 6-12 months using one or more imaging modalities: TTE (8/8), MRI (6/8), TEE (2/8). There was no consensus on optimal timing of cardiac catherization to measure pulmonary artery pressures (PAP), necessary in the decision-making with respect to intervention for sTR, cardiac transplantation and/or mechanical assistance.
Conclusion: Almost 30000 adult congenital heart disease patients are actively followed across Canada in specialized multidisciplinary clinics including over 520 Mustard and 90 Senning patients. There appears to be consensus amongst Canadian congenital cardiac surgeons on the modalities and follow up timing of Mustard and Senning adult patients but not on the

P2950 - TRANSPOSITION OF THE GREAT ARTERIES ARTERIAL SWITCH IN A 32 YEARS OLD FEMALE CASE REPORT

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The transposition of the great arteries is a congenital heart malformation characterized for an atrioventricular concordance and a ventriculoarterial discordance. It has an incidence of 1 in 3500-5000 newborns. The association with other heart malformations, like a ventricular septal defect or a left ventricular outflow obstruction, determine the time and the form of clinical presentation. It is generally a lethal congenital malformation, with a life expectancy of approximately 9 months in non-operated patients. We present the case of a 32-year-old female patient with a diagnosis of D Transposition of the large arteries with pulmonary stenosis and atrial and ventricular septal defects, which was taken to surgical anatomical correction by performing an arterial switch (Jatene procedure) with an extension of the neaorta valvular

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annulus (Nicks) and mechanical valvular prosthesis placement, as well as closure of the septal defects, with a favorable evolution.

P2984 - A CASE OF CONGENITAL LEFT CORONARY ARTERY ATRESIA AND MITRAL INSUFFICIENCY

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Left main coronary artery atresia is a rare coronary anomaly in which there is no left coronary ostium,blood flows from the right coronary artery to the left through small collateral arteries and retrogradely. Since published case reports are few and rather scattered. The case of a 44years-old men with atresia of the left main coronary ostium and mitral insufficiency is presented. The men underwent a left internal thoracic-left anterior descending coronary artery bypass and plasty of the mitral valve. A selective study of the left internal thoracic artery demonstrated excellent flow into the left coronary artery system. In view of the occurrence of sudden death and massive myocardial infarction in adult patients shown to have atresia of the left main coronary artery, it is suggested that adult patients with this condition, who require open-heart surgery for any other cardiac disorder, should undergo coronary artery bypass grafting concurrently.

P3047 - EXERCISE CAPACITY AFTER PERCUTANEOUS BALLOON VALVULOPLASTY FOR CONGENITAL PULMONARY STENOSIS IN LONG TERM FOLLOW UP

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Medical University of Gdańsk, Department of Paediatric Cardiology and Congenital Heart Diseases, Poland-Poland

Introduction: Early studies have shown congenital valvular pulmonary stenosis (PS) treated by percutaneous balloon pulmonary valvuloplasty (PBPV) to have excellent outcomes with respect to relief of PS and risk of pulmonary regurgitation (PR) provided proper balloon diameter was selected. However, more recent long-term studies have shown that patients may have suboptimal functional outcome, which may be related to residual changes, mainly PR. The aim of this study was to assess exercise capacity by Bruce protocol in long-term survivors of PBPV.
Methods: Fifty-six patients treated by PBPV underwent detailed medical and echocardiographic assessment. Those without contraindications underwent a treadmill stress-test in accordance with maximal exercise Bruce protocol. Exercise capacity index (ECI) was calculated as z-score based on age- and sex-relevant normative data. Data is presented as mean ± standard deviation or median (range) dependending on distribution.
Results: Median time of observation was 13,6 (5,7–24,2) years. Eleven patients were excluded from stress-test due to contraindications – handicap (n = 5), other cardiac disease not related to PS nor PBPV (n = 3), orthopaedic (n = 2), infection (n = 1). The remaining 45 patients were 56% female and 20,5 (6,8–48,4) years of age. The exercise time was 10,9 ± 2,2 minutes and METs was 11,4 ± 1,8 kcal/kg*h), ECI was 0,1 ± 0,9 (-2,2–3,1). All patients had normal HR and BP response and terminated exercise due to symptoms of exertion. Only 6% (n = 3) of patients had a significantly reduced ECI and in 11% (n = 5) ECI reduction was mild. Further analysis indicated that the reduction was correlated with comorbidities (e.g. asthma, obesity) rather than residual transvalvular gradient or pulmonary regurgitation.
Conclusion: There is no overt reduction in exercise tolerance related to outcome of PS treatment by PBPV at long-term follow-up. This pilot study is limited by a small number of patients and use of exercise time or METs as exercise capacity proxy.

AMBULATORY

P1080 - THE LONG TERM FOLLOW UP OF FUNCTIONAL HEALTH STATUS OF ADOLESCENTS WHO HAVE UNDERGONE CARDIOPULMONARY BYPASS IN INFANCY

Wotherspoon Jane[1], Eagleson Karen [2], Auld Ben[2], Johnson Susan[2], Stocker Christian[2], Heussler Honey[3], Ware Robert[4], Gilmore Linda[5], Justo Robert[2]
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Background: Congenital heart surgery survivors are at risk of poorer functional health outcomes compared with healthy peers. 35 neurologically normal infants aged less than 6 months underwent cardiopulmonary bypass surgery for congenital heart disease between 1999 and 2001. Assessment at 1 and 5 years post procedure demonstrated impaired cognitive, behavioural and neuropsychological performance. This study describes the neuropsychological and functional health of adolescents with congenital heart disease followed since infancy.
Methods: This was a prospective cohort study. Participants underwent medical review, genetic screening, health-related quality of life (HRQoL) assessment and a comprehensive battery of neuropsychological tests. Parents also completed rating scales.
Results: 21 patients, 95% of whom had a biventricular circulation, were assessed at median age 15.4 years (range, 14–17 years). All patients were NYHA classification 1, reporting peak physical activity ranging from sedentary to competitive sport. A deletion was detected on 8q22.3 following SNP array analysis in one patient. While global intelligence was in the average range (94.3 + 12.7; p = 0.06), adolescents scored significantly lower than expected across various neurodevelopmental areas compared to the general population including cognitive domains of working memory and processing speed. Participants also performed below expected on a test of mathematical achievement. Deficits in attention, executive function and visual-spatial ability were found. In social-emotional and adaptive domains, participants reported more learning problems, poorer attitude to teachers, higher sense of inadequacy and poorer self-reliance. Parent scales indicated poorer home living abilities in their children. Adolescents described more positive family relations, while parents reported less aggression and fewer externalising problems. Adolescents and parents reported significantly lower HRQoL across all domains.
Conclusions: These results are consistent with previously described outcomes for this cohort, and with patterns of impairment reported in international studies. The importance of early intervention is emphasised, as early deficits extend into young adulthood.

P1120 - CHARACTERISTICS OF ADMISSIONS FOR KAWASAKI DISEASE FROM 1997 TO 2012 LESSONS FROM A NATIONAL DATABASE

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Background: Kawasaki Disease (KD) is one of the most common causes of acquired heart disease in children. While countries like Japan have characterized national characteristics of KD, such data has not yet been presented for cases in the United States. The aim of this study was to characterize Kawasaki Disease admission using data from a national database.
Materials and Methods: Data from the Kids' Inpatient Database from 1997 to 2012 was used for this cross-sectional study. Characteristics of KD admissions as well as those without KD were collected/ Multivariate analysis was done to identify risk factors associated with KD. Multivariate analysis was also conducted to determine whether KD is an independent risk factor for cardiogenic shock or coronary artery aneurysm.
Results: A total of 15,962,403 admissions were included in this analysis. Of these, 17,918 (0.11%) were for KD. For those with KD, the median age of admission was 2 years with a (range 1 month to 20 years). The median length of admission was 3 days (range 1 to 76 days). Median cost of hospitalization was 15,186 US dollars. Of those with KD admission, 485 (2.7%) were found to have coronary artery aneurysms, 19 (0.1%) suffered from cardiogenic shock, and 5 (0.1%) required extracorporeal membrane oxygenation. Only 4 patients (0.01%) experienced inpatient mortality. KD was not an independent risk factor for cardiogenic shock or need for extracorporeal membrane oxygenation. KD was an independent risk factor for coronary artery aneurysm (odds ratio 2,882.381, 95% confidence interval 1,848.024 to 3,395.679).
Conclusion: KD accounts for a small percent of total pediatric inpatient admissons but is associated with high cost of hospitalization. Certain races are associated with an increased risk of KD as is younger age. KD is an independent risk factor for coronary artery aneurysms bot not cardiogenic shock.

P1127 - RISK FACTORS ASSOCIATED WITH RECURRENCE OF KAWASAKI DISEASE IN MEXICAN CHILDREN

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Background: Kawasaki disease (KD) is an acute febrile illness characterized by systemic vasculitis. Recurrences of KD (defined as at least three clinical signs of KD in addition to fever ≥ 5 days), presenting ≥ 14 days after the return to baseline from the index episode is reported in approximately 3-4% of all cases in Japan.
Objective: To assess the frequency and determined the risk factors associated with recurrences of KD in patients treated at the Instituto Nacional de Pediatría in Mexico City.
Material and Methods: An observational, comparative, retrospective and case-control study of all patients diagnosed with recurrent KD in our Institution from August 1st, 1995 to August 31st, 2016 was performed. The clinical presentation, laboratory results, treatment used and coronary artery abnormalities in the recurrent-KD and non-recurrent KD groups were analyzed and compared.
Results: We included 525 patients with KD diagnosed at our institution; we had 21 recurrences of KD (4.0%), 17 patients had one recurrence, 1 patient had 2 recurrences and 1 patient had 3 recurrences of KD. 19 cases or our cases were male (90.4%) with mean age at diagnosis of the first episode of 29.81 ± 35.10 months and with a mean of 15.42 months of the new event after the initial episode (1 to 60 mo.) In bivariate analysis, male gender ($p < 0.037$), central nervous system manifestations in the acute phase of KD ($p < 0.053$) and coronary aneurysms at diagnosis ($p < 0.05$) showed statistical significance. There were no factors associated with recurrence in a multivariate analysis.
Conclusions: This is a very small series of KD with a slightly increased rate of recurrence compared with the rate of recurrences reported in the literature. But to allow an early recognition of a new event, a previous history of KD should be considered to initiate.

P1141 - MANAGEMENT DIFFICULTY OF LATE DIAGNOSED TRANSPOSITIONS OF THE GREAT VESSELS IN A DEVELOPING COUNTRY

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Background: Transposition of the great vessels (TGV) is a relatively frequent congenital cyanotic heart disease. The diagnosis is made in the antenatal or neonatal period and surgical correction should be rapid, usually at the age of 15 days. However, in our country, this heart disease is belatedly diagnosed and thus belatedly operated. Objective: Describe the clinical features and the management difficulties of late-diagnosed TGVs.
Methods: We conducted a retrospective observationnel study in the department of Pediatrics of Sahloul hospital-Sousse. All children with late-diagnosed TGV were included. The study period went from 2000 until 2015.
Results: We collected 10 cases of TGV (4 girls, 6 boys). The average age of diagnosis was 22 days. The clinical features at admission were a cyanosis in 100% of cases associated with heart failure signs at 50% of patients with hemodynamic instability requiring mechanical ventilation. TGV was isolated in 2 cases, associated with CIV in 7 cases; and in one case, it was associated with pulmonary stenosis, aortic coarctation and hypoplasia of the aorta. A Rashkind was done in 6 cases within 6 days. The arterial Switch was performed in 5 cases after 31 days on average. Only one patient was placed on ECMO after surgery and no child had pulmonary banding before doing the Switch. The outcome of these patients was favorable in 4 cases; 1 patient died secondary to Septic shock, 2 months after surgery. Patients who not underwent surgery (5 cases), died within 30 days on average.
Conclusion: Children with TGV require urgent diagnosis and management. High rates of morbidity and perioperative mortality of late-diagnosed TGV result of the inability of the right ventricle to ensure systemic circulation. This situation may begin even before the age of 1 month as illustrated by our study.

P1158 - CLINICAL IMPORTANCE OF SCHOOL CARDIAC SCREENING WITH UNIVERSAL ELECTROCARIOGRAPHIC SCREENING IN THE DIAGNOSIS OF ASYMPTOMATIC YOUNG PATIENTS WITH ATRIAL SEPTAL DEFECT

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Background: Japanese government has enforced a law of regular school cardiac screening (SCS) with universal electrocardiographic (ECG) screening at 1st, 7th, and 10th grader in 1995. The aim of

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this study was to determine impact of SCS in the management of patients with atrial septal defect (ASD).
Methods: Subjects were 253 patients with ASD aged between 6.5 and 35 years old that had documented reasons why patients were sent to cardiologists. Based on the retrospective chart review including demographics, past history, physical examination and ECG findings at SCS, we determined the role of SCS in the diagnosis of ASD.
Results: About half of patients (48%) were detected at SCS and the remaining patients were diagnosed at younger age mainly because of cardiac murmur (87%). In patients detected at SCS, three fourths (75%) of patients were identified by ECG screening due to incomplete RBBB with right axis deviation or incomplete RBBB with rR' pattern and the remaining one fourth patients were mainly identified by cardiac murmur. Notably, 60% of these patients identified by ECG abnormality at SCS did not have significant cardiac murmur, >= 2/6 in Levine grading. There was no significant difference in age (13.8 ± 2.5 vs. 14.5 ± 2.9 years old), Qp/Qs (2.5 ± 1.0 vs. 2.4 ± 0.9), and mean PAP 14 ± 3 vs. 15 ± 3 mmHg) between patients with and without significant cardiac murmur. As summary, 22% of patients with asymptomatic ASD in this age group might not be diagnosed if there is no ECG screening in Japan.
Conclusions: SCS, especially combined with universal ECG screening, has significant role to identify asymptomatic patients with ASD in Japan though cost effectiveness must be determined.

P1164 - KAWASAKI DISEASE IN CHILDREN YOUNGER THAN 1 YEAR OF AGE IN A MEXICAN POPULATION

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Background: Kawasaki disease (KD) is an acute febrile vasculitis of unknown origin. KD represents the most common cause of acquired heart disease in children in developed countries. KD is uncommon in patients younger than 1 year of age and in Mexico there are few reports of KD in this group of age.
Objective: To describe the clinical features, laboratory parameters, treatment employed and outcome of patients with KD younger than 1 year of age in a third level facility in Mexico City.
Methods: A cross-sectional study was conducted at the Instituto Nacional de Pediatria in Mexico City, Mexico From August 1995 to August 2016. Children with KD younger than 12 months of age were used as the case group and patients older than 12 months of age as a control group. P < 005 was considered to be statistically significant.
Results: During the study period, 519 patients were diagnosed with KD at our Institution; 116 patients (22.3%) were younger than 1 year of age. The mean age at diagnosis was 8.16 ± 2.95 months. Seventy-nine patients were male (68.10%). Mean time from the onset of fever to diagnosis of KD was 10.15 ± 6.852 days (range 3 to 41 days). An incomplete form of KD was present in 34 patients (29.31%). Fifty-three of our patients developed coronary aneurysms (z-score >2.5), and seventeen of them were categorized as giant aneurysms; two of these patients died of cardiogenic shock in the acute phase of the disease.
Conclusions: In Latin American countries are few reports of KD in patients younger than 1 year of age. In our experience, more patients had coronary artery aneurysms than the reported in the literature.

P1166 - KAWASAKI DISEASE IN PATIENTS OLDER THAN 8 YEARS OLD IN A CHILDREN'S HOSPITAL IN MEXICO CITY. IS THERE REALLY A DIFFERENCE IN PRESENTATION AND OUTCOME

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Background: Kawasaki Disease (KD) is an acute febrile illness characterized by systemic vasculitis of unknown etiology. Cardiac sequelae, such as coronary artery aneurysms (CAA), are one of the most important aspects of this disease. KD is rarely diagnosed in older children.
Objective: To describe the clinical and laboratory features, cardiac sequelae and outcome in children older than 8-years old with KD in a third-level facility in Mexico City.
Methods: A cross-sectional and comparative study was conducted at the Instituto Nacional de Pediatria in Mexico City, Mexico From August 1995 to August 2016. Children with KD older than 8 years were used as the case group and patients younger than 8 years were used as the control group. $p < 0.05$ was considered to be statistically significant.
Results: 519 cases of KD were diagnosed during the study period, 37 cases of them were older than 8-years old, (7.12%), with a mean age of 137.92 ± 31.24 months (range 96 to 200 months). There were no statistically difference in the classical KD signs, but older children more often had arthralgias ($p < 0.015$) congestive heart failure ($p < 0.020$) and an incomplete form of KD ($p < 0.034$). We found no difference in the development of coronary artery aneurysms in both groups. Almost all patients in both groups received IVIG but patients in the older group required a second IVIG dose ($p < 0.01$)
Conclusions: KD in patients older than 8-years old represent a clinical challenge because in most of the cases they presented with an atypical clinical picture, which contribute to a delayed diagnosis.

P1175 - GRADUAL DETERIORATION OF VITAMIN D STATUS AND BONE MASS DENSITY IN CHILDREN AND ADOLESCENTS WITH FONTAN CIRCULATION

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Background: Our group has shown high prevalence of vitamin D deficiency and low bone mass density in adolescents with Fontan circulation. The study aim was to investigate vitamin D status and bone mass density in children with Fontan circulation at different ages.
Material and Methods: Patients were recruited from our national Fontan clinic and divided in two cohorts aged 5 to 9 years and 10 to 15 years respectively. We prospectively measured serum levels of 25-OH-vitamin D and total body bone mass density by narrow

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fan-beam dual energy x-ray absorptiometry (DXA) scan. The DXA results were compared to age and gender matched reference data and expressed as Z-scores. Vitamin D and DXA results of the two cohorts were compared to the previous cohort of 17 patients aged 16 to 18 years. Current recommendations for patients with chronic illness suggest an optimal serum level of 75 – 125 nmol/L.
Results: We included 15 patients aged 5 to 9 years and 19 patients aged 10 to 15 years. Vitamin D levels in the two cohorts were 77.5 ± 31.0 nmol/L (mean ± SD) and 61.6 ± 30.2 nmol/L respectively, while the level in the adolescent cohort was 34.6 ± 18.3 nmol/L. The DXA scans for the two youngest cohorts showed z-scores of 0.7 ± 0.9 and -0.7 ± 1.4, while the oldest cohort had a z-score of -0.8 ± 1.1. Apart from BMD in age group 2 and 3 ($p = 0.74$), all group differences were statistically significant.
Conclusion: Both vitamin D status and bone mass density show age-related differences in Fontan patients and a significant decrease through childhood and adolescence. Whether this is due to life style factors (i.e. sun exposure, dietary intake of vitamin D and physical activity) or due to a pathophysiological effect of the Fontan circulation is unknown.

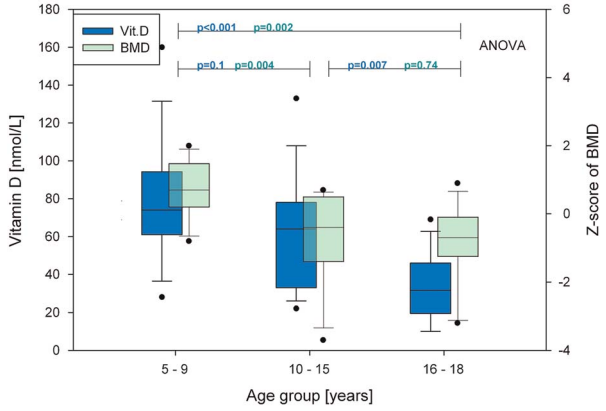

Figure.

P1191 - RISK FACTORS FOR DEVELOPMENT OF CORONARY ARTERY ANEURYSMS IN KAWASAKI DISEASE IN MEXICAN CHILDREN

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Background: Kawasaki Disease (KD) is an acute febrile illness characterized by systemic vasculitis of unknown etiology. Coronary artery aneurysms (CAA), are one of the most important complications of the disease. Actually, it is the major cause of acquired heart disease in developed countries but its frequency in Mexico is still unknown.
Objective: To establish the risk factors for development of coronary artery lesions in children with KD who were treated at the Instituto Nacional de Pediatria in Mexico City.
Material and Methods: An observational, comparative, retrospective case-control study of all patients diagnosed with KD in our Institution from August 1995 to August 2016. We reviewed the medical records and analyzed gender, age, weight, height, clinical manifestations, time from the onset of the symptoms to diagnosis, hemoglobin, leucocyte count, platelet count, ESR, C-RP, albumin, sodium, AST ASL, treatment used and the development of coronary artery aneurysms.
Results: We studied 499 cases of KD, 65.9% were male with a mean age at diagnosis of 37.87 ± 35.81 months. The mean duration of fever from the onset of the symptoms to diagnosis was 9.1 ± 548. An incomplete form of KD was diagnosed in 103 patients (20.64%). 165 patients developed CAA (33%). Multivariate analysis for CAA showed that younger age ($p < 0.004$) prolonged time to diagnosis ($p < 0.001$), central nervous system manifestations ($p < 0.008$) anemia ($p < 0.001$) leukocytosis ($p < 0.005$), thrombocytosis ($p < 0.041$) were the most important risk factors for development of CAA in our patients.
Conclusions: This is the largest series of KD in Mexico, with most cases diagnosed in recent years, so it appear that the disease is more common than initially thought. Also, the frequency of CAA is greater than the reported in the literature, but it appears to be related to a delayed

P1196 - CONTROVERSIES IN ASSOCIATION OF CARDIORESPIRATORY FITNESS AND ARTERIAL STIFFNESS IN CHILDREN AND ADOLESCENTS

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Objective: The relationship of cardiorespiratory fitness and arterial compliance in children and adolescents remains controversial. The aim of this study was to assess this association with a quantitative approach.
Patients and Methods: 646 healthy school children and adolescents (316 female, age 13.9 ± 2.1 years) were cross-sectionally investigated in seven school settings in and around Munich for their cardiorespiratory fitness, demographic, anthropometric and hemodynamic parameters. Surrogates of arterial stiffness like pulse wave velocity (PWV), augmentation index normalized to a heart rate of 75 (AI@75), peripheral and central systolic blood pressures were measured in a supine position using the oscilliometric Mobil-O-Graph®. Cardiopulmonary fitness was measured with a six minute indoor run tests.
Results: After correction for age, sex, body mass, body height, heart rate and mean arterial pressure controversial findings occurred. PWV increased with higher cardiorespiratory fitness (Beta = .173; $p < .001$) which means an unfavorable relation, whereas AI@75 declined with higher cardiorespiratory fitness (Beta = -.106; $p = .025$). Higher cardiorespiratory fitness seems therefore, in contrast to PWV, beneficial for AI@75. The third surrogate of arterial stiffness, central systolic blood pressure, showed no association to cardiorespiratory fitness (Beta = .066; $p = .052$).
Conclusions: Different surrogates of arterial stiffness provide different associations to cardiorespiratory fitness in children and adolescents still after correction for multiple confounder. More research is needed in this field to understand functioning of the juvenile vessels and also measurements and methodological approaches should be reconsidered.

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P1203 - SHORT STATURE PREVALENCE BEFORE CARDIAC SURGERY IN CHILDREN WITH CONGENITAL HEART DISEASE

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Background: Children with congenital heart disease (CHD) frequently present malnutrition, being a possible long-term consequence short stature (SS). The aim of this study was to compare the nutritional status and prevalence of SS in children according to CHD prior to surgical intervention.
Methods: Retrospective study in a South American cardio-surgical center. We studied children with CHD undergoing surgery with extracorporeal circulation between 2009 and 2013. Patients with previous history of prematurity, genotype or other illness with possible nutritional compromise were excluded. An anthropometric evaluation was performed per Z-score and World Health Organization standards. A comparison of SS prevalence according to CHD was conducted.
Results: Six hundred and forty-three cardiac surgeries with cardiopulmonary bypass were performed during the period in 549 patients, 55.7% men (median age: 9.7, interquartile range 3.3-39.2 months). The mean height-for-age Z-score (HAZ) was -1.0 ± 1.3 and 21.5% of the patients presented SS (HAZ < -2). The mean weight-for-height Z-score (WHZ) was -0.3 ± 1.5, 11.7% of the patients presented wasting (WHZ < -2) and 5.8% obesity (WHZ $> +2$). Patients with ventricular septal defect (VSD), and tetralogy of Fallot (TOF) presented the highest SS prevalence (30.3% and 31.3%, respectively) versus in atrial septal defect with the lowest SS prevalence, 17% ($p < 0.01$). In patients with univentricular physiology undergoing to Fontan circulation, 4 of 44 patients (9.1%) presented SS before the first stage (mean HAZ -0.7 ± 1.3), 14 of 47 (29.8%) prior to the second stage (HAZ -1.5 ± 1.1) and 3 of 23 (13%) prior to the Fontan (HAZ -0.4 ± 1.0, $p < 0.01$ between stages). No correlation was found between the age of surgery and HAZ.
Conclusions: There is a high prevalence of SS in patients prior to CHD surgery, especially in patients with VSD, TOF and prior to the second stage. As SS is an indicator of chronic malnutrition, it is relevant to optimize nutritional support in these patients.

P1204 - COMPARISON OF THE EFFECTIVENESS OF FOUR(4) SESSION PLAY MODULE VERSUS THE CONVENTIONAL CARDIOVASCULAR REHABILITATION THERAPY AMONG POST OPERATIVE CONGENITAL HEART DISEASE PEDIATRIC PATIENTS AGES 5 12 YEARS OLD

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Background: Play is primarily a way for a child to communicate, especially when under stress. However, the efficacy of therapeutic play is not widely accepted by the scientific community due to lack of evidence-based studies. The objective of this study was to determine the effectiveness of the four-session play module as cardiovascular post-operative rehabilitation therapy.
Methodology: A randomized controlled clinical trial among pediatric patients ages 5-12 years old diagnosed of Congenital Heart Disease, who underwent elective open heart surgery. The patients were divided into two groups: play module or conventional rehabilitation therapy. Quality of life (QoL) was assessed through PedsQL cardiac module.
Result: There was a total of 56 subjects, with a mean age of 7.3 years. On the 2nd post-operative day, there was a significant difference on the QoL of patients who had the play module with regards to heart problem (2.4 ± 0.77 vs 1.4 ± 0.74, $p < 0.001$), treatment problem (1.2 ± 1.12 vs 0.5 ± 0.74, $p = 0.019$), perceived physical appearance (2.2 ± 1.09 vs 1.4 ± 1.1, $p = 0.007$), treatment anxiety (2.6 ± 0.99 vs 1.4 ± 1.14, $p < 0.001$), cognitive (1.7 ± 0.94 vs 0.8 ± 0.69, $p < 0.001$), and communication (2.4 ± 1.15 vs 1.3 ± 1.1, $p < 0.001$). The QoL was also significant on the 5th post-operative day, except for treatment problem. Comparing the treatment group, there was a significant improvement on the QoL as the sessions progressed.
Conclusion: The structured play module is an effective therapeutic tool for cardiovascular rehabilitation of post-operative pediatric patients. It helped improved the quality of their lives post-operatively.

P1218 - MUSCULAR ENDURANCE AND AGILITY LIMIT THE PHYSICAL ACTIVITY CAPACITY OF CHILDREN WITH CONGENITAL HEART DEFECTS COMPARED TO CANADIAN SCHOOLCHILDREN

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Background/Hypothesis: Physical literacy is the motivation, confidence, knowledge and physical competence to pursue physical activity for life. Children with congenital heart defects (CHD) are more sedentary than peers, suggesting a physical literacy deficit. It was hypothesized that children with CHD would demonstrate physical competence and daily behavior deficits.
Materials and Methods: Cross-sectional study of children, 8 to 12 years of age, with CHD ($n = 37$, 35% female) or attending local schools ($n = 963$, 55% female). The Canadian Assessment of Physical Literacy assessed total physical literacy and four subdomains (daily behaviour, physical competence, motivation and confidence, and knowledge and understanding).
Results: Children with CHD walked more steps each day ($13{,}750 \pm 3457$; $p < 0.001$) and tended to report more sedentary behaviour ($p = 0.02$), as shown in Table 1. Their muscular endurance was only 60% of that achieved by school children of similar age (CHD: 3.6 ± 2.4; School: 6.0 ± 4.4; $p < 0.001$). CHD children also tended to have lower agility scores ($p = 0.02$). In multi-variable regression models accounting for age, sex, and season of assessment, higher physical competence scores occurred among school children, older children, males, and those tested during the summer (model $R2 = 0.13$). Age, sex, season and study group explained very little of the variance in physical literacy scores (model $R^2 \leq 0.03$).
Conclusions: Children with CHD have motivation for, and knowledge about physical activity that is similar to peers. However, muscular endurance and agility were decreased even among our highly active sample of CHD patients. The impact of interventions to enhance muscular endurance and agility and the

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physical literacy of a randomly selected sample of children with CHD should be pursued.

Table 1. Physical Literacy of Children with Congenital Heart Defects Compared to Local Schoolchildren

CHD patient and school children differences[1]	CHD[2] (mean ± SD)	School (mean ± SD)	99% Conf. Int.[3]	p-value
SimilarTotal Physical Literacy Score	63.7 ± 9.7	62.3 ± 10.1	-6.0,3.4	0.46
Similar Daily Behavior	20.3 ± 5.9	18.3 ± 6.5	-4.9,0.9	0.07
CHDwalked more steps each day	13750 ± 3457	11294 ± 3259	**811,4100**	**< 0.001**
Similar self-reported sedentary behavior	3.6 ± 3.2	5.0 ± 2.9	*-0.09,2.77*	*0.02*
Similar Physical Competence	18.3 ± 3.7	19.7 ± 3.8	*-0.42,3.06*	*0.05*
Similar agility and movement skill	30.3 ± 7.0	33.0 ± 5.4	*-0.42,5.89*	*0.02*
Similar endurance(PACER shuttle run)	17.0 ± 9.4	17.3 ± 9.2	-4.00,4.52	0.87
Similar strength (grip strength)	5.0 ± 4.6	5.7 ± 4.8	-1.41,2.80	0.38
Similar flexibility (sit and reach)	4.2 ± 2.1	4.7 ± 2.4	-0.52,1.45	0.21
Similar body composition	31.2 ± 3.3	31.0 ± 5.8	-3.00,2.44	0.83
Similar waist circumference	16.0 ± 3.0	15.9 ± 3.0	-1.51,1.22	0.77
CHD had lower muscular endurance	3.6 ± 2.4	6.0 ± 4.4	**1.24,3.53**	**< 0.001**
Similar Motivation and Confidence	12.2 ± 3.1	12.4 ± 2.7	-1.26,1.57	0.77
Similar Knowledge and Understanding	11.8 ± 2.7	11.5 ± 2.6	-1.57,0.91	0.48

[1] For all measures a higher score represents a healthier behavior (e.g., higher sedentary score = less sedentary time).
[2] CHD = congenital heart defect; School = children tested in local schools.
[3] Conf. Int. = confidence interval; significance set at $p < 0.01$ to account for multiple outcomes ($p = 0.05$ / 5 primary outcomes (total + 4 domain scores)); significant differences shown in bold; *trends shown in italics*.

P1255 - BICUSPID AORTIC VALVE IN CHILDREN WITH MARFAN SYNDROME HIGH APPEARANCE WITHOUT CORRELATION TO SEVERITY OF CLINICAL SYMPTOMS

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Background: Due to age dependent development of most pathologies in Marfan syndrome (MFS) estimation of clinical outcome in childhood is difficult. Bicuspid aortic valve (BAV) is correlated with MFS and prevalence is up to 4.7% in adult Marfan patients. Data in childhood about appearance and correlation with severity of phenotype is missing. Subsequently it may be a predictor for onset of pathologies and helpful to determine individual patient care. We hereby evaluated correlation of present BAV with Marfan pathologies in childhood.
Material and Methods: Since 2008 we investigated 395 patients (11.4 ± 5.5 y) with suspected MFS. In 145 patients MFS was diagnosed, thereby 16 patients showed BAV. We retrospectively analyzed correlation of prevalence and age of onset of cardiovascular pathologies (Dilatation sinus of valsalvae (SV), Mitral valve prolaps (MVP)), systemic manifestation of Ghent Criteria and FBN1 mutation with appearance of BAV.
Results: Prevalence of BAV in pediatric Marfan patients was 11.0%. Age of patients with and without BAV did not differ significantly. There is no correlation of BAV with prevalence or age of onset of marfan pathologies (SV, MVP, systemic manifestation, ectopia lentis, FBN1 mutation) of MFS ($p > 0.05$).
Conclusions: In our large collective prevalence of BAV in MFS was surprisingly higher compared to adult. The reason is unknown, but can possibly be an effect of selection. Indeed, occurrence of BAV did not correlate with severity of clinical phenotype. Especially SV dilatation did not appear earlier or more often in BAV patients. Thus BAV is present frequently in MFS but is no predictor for clinical outcome and estimation of need for therapy. Development of pathologies in those patients in adulthood is unknown but may be relevant for course of disease. Good predictors for severity of phenotype in childhood are unfortunately still missing.

P1273 - EARLY FUNCTIONAL HEALTH OUTCOMES FOR INFANTS AND THEIR FAMILIES WHO HAVE UNDERGONE ARTERIAL SWITCH OR NORWOOD PROCEDURES

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Background: This Program assessed the impact on functional health, in the current era, of cardiac bypass surgery on a cohort of neonates at risk of neurodevelopmental disability.
Methods: Infants undergoing Arterial Switch Operation (ASO) or Norwood procedure from January 2013 were enrolled prospectively into the Neurodevelopmental Program. Demographic and clinical data were analysed. Bayley Scales of Development Third Edition, Health-Related Quality of Life (HRQoL) and Parenting Stress assessments were performed at 6, 12 and 24 months.
Results: ASO patients had median Comprehensive Aristotle Complexity Scores (CACS) of 12 (IQR 10 – 14) and intensive care (PICU) stay of 8.8 days (IQR 6.0-11.9). Norwood infants had median CACS of 17 (IQR 14 – 20) and PICU stay of 21.4 days (IQR 16.1-27.1). Infants were assessed at 6 (ASO: n = 25, Norwood: n = 13), 12 (ASO: n = 20, Norwood: n = 8) and 24 months (ASO: n = 7, Norwood: n = 3). ASO infants scored in low average range in adaptive behaviour at 12 months (86.9 ± 24.5), and at least average range across other domains at 6, 12 and 24 months. Norwood infants scored in low average range in adaptive behaviour but average range across all domains at 6 months. Subsequently they scored in low average-borderline range; in language (12 month: 83.6 ± 35.9, 24 month: 85.3 ± 33.2), motor (12 month: 71.9 ±29.8, 24 month: 86.3 ± 21.5) and adaptive behaviour (6 month: 88.64 ± 30.4, 12 month: 85.9 ± 12.4, 24 month: 77.7 ± 23.0) domains. Parent-reported total HRQoL was significantly lower in ASO group at 6 months only. Lower total HRQoL in the Norwood group was persistent at 12 months. Parenting stress was in the normal range for both groups at all time point.
Conclusion: Study findings suggest that infants undergoing the Norwood procedure have reduced functional health when followed beyond 12 months.

P1293 - THE MENTAL HEALTH OF ADOLESCENTS LIVING WITH POTENTIALLY FATAL ARRHYTHMIA A SYSTEMATIC REVIEW OF THE LITERATURE

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Background/Hypothesis: Potentially fatal arrhythmia diagnoses add to the mental health challenges of adolescence. This systematic review sought to summarise current knowledge regarding the mental health of adolescents diagnosed with a life-threatening cardiac arrhythmia.
Materials and Methods: Search terms using psychological problems of environmental origin and inherited cardiac arrhythmia diagnoses identified studies among those 9 to 18 years. Multiple authors screened 3238 titles/abstracts and reviewed citations to identify 233 potentially relevant articles. Full text review identified 15 articles meeting inclusion criteria.
Results: All 15 studies (see table) were cross-sectional; 8/15 required an implantable cardioverter defibrillator (ICD). Methods were quantitative (n = 10), qualitative (n = 4) or mixed (n = 1), with 16% to 100% of participants having an inherited cardiac diagnosis. Patients and parents reported lower quality of life, particularly in relation to physical function, social relationship difficulties, being restricted from peer activities, bodily pain and mental and emotional health. Mental health studies (n = 8) suggest that mental and emotional health issues are not extensive among patients with ICDs assessed 2 + years after implant. Children's self-perceptions and behaviour were similar to healthy populations and rates of anxiety and depression were not increased. However, psychosocial stresses may be more significant in the months immediately after diagnosis. Anxiety, depression or aggressive behaviour is reported by 15-33% of these patients, with higher risk among patients who also have a diagnosed sibling, those with cardiomyopathy, and those who report decreased quality of life.
Conclusions: Mental health research among adolescents with inherited arrhythmias is extremely limited and of low quality. Data, primarily from patients 2-4 years post ICD implant or diagnosis, indicate quality of life may be decreased and 15-33% experience mental health issues. The mental health of adolescents with inherited arrhythmias, whether or not they have an ICD and from time of diagnosis, and the impact of clinical interventions remain unknown.

Table. Description of Study Design and Participants.

Citation	Study Design	N (total)	N (genetic cardiac)	Research Method
Antiel et al, 2016 USA	Cross-Sectional Quantitative	26/62 participants were 5-18 yrs 3.5 ± 2.1 years after left ventricle denervation	23 (89%) 17 LQTS, 6CPVT	CHQ-PF28 CSDS
Bratt et al, 2012 Sweden	Cross-Sectional Qualitative	13 asymptomatic HCM youth (median age at diagnosis 11 yrs) 12-24 months post-diagnosis	13(100%) 13 HCM (asymptomatic)	Interview
Costa et al, 2007 Brazil	Cross-Sectional Mixed	15 youth (13 to 21 years) with ICD 2.6 ±1.8 years after implant	13(72%) 13 to 18 years 5 HCM, 2 DCM, 1 ARVC, 2 CPVT, 2 LQTS, 1 Brugada	Interview SF-36 Survey
Czosek et al, 2016 USA	Cross-Sectional Quantitative	61 patient-parent pairs from 45 families Compared to healthy published norms and CHD cohorts (BAV.TOF, Fontan) 8(13%) with device Dx: 4 ± 3yrs	61 (100%) 61 LQTS	PCQLI PedsQL Self-Perception Profile Achenbach Youth Self-Report & Parent CBCL
Czosek et al, 2015 USA & UK	Retrospective Quantitative	288 patient-parent pairs 53 CCHB, 27 VT, 90 SVT, 118 LQTS,	118(14%)118 LQTS	PCQLI
Czosek et al, 2012 USA	Cross-Sectional Quantitative	173 patient-parent pairs with ICD (n = 40, Dx: 5.5 ± 4.4 yrs) or pacemaker (n = 133. Dx: 10.8 ± 3.9)	27(16%) 11 HCM, 2 DCM, 14 LQTS	PedsQL PCQLI CBC Self-Perception Profile
Demaso et al, 2004 USA	Cross-Sectional Quantitative	20 patient-parent pairs with ICD 1.4 years post implant (median 0.1, range 0 to 6)	4 sustained polymorphic VT (20%)	Revised Manifest Anxiety Reynold's Depression CHQ-87/50 Worries About ICD Scale
Eicken et al, 2006 Germany	Cross-Sectional Quantitative	16 patients with ICD 43.1 months (range 1-105 months) post implant	9 (56%) 2 HCM, 7 electrical	DISYPS-KJ
Giuffre et al, 2008 Canada	Cross-Sectional Quantitative	47 patient/mother pairs with asthma or LQTS Dx: Not specified	7(15%) LQTS	Fear Survey Schedule Revised Manifest Anxiety Achenbach CBCL State-Trait Anxiety
Koopman et al, 2012 Netherlands	Cross-Sectional Quantitative	30 patients with ICD 16.3 years (range 9-23) 3.6 years post implant	23 (77%) 6 cardiomyopathy 17 primary electrical	Symptom Checklist (SCL-90- R) Worries about ICDs Scale Parent for demographics
Meulenkamp et al, 2008 Netherlands	Cross-Sectional Qualitative	33 patients with genetic cardiovascular disease ≥ 6 months post Dx	17(52%)11 LQTS, 6 HCM	Interview
Rahman et al, 2011 Australia	Cross-Sectional Qualitative	6 patient-parent pairs with ICDs 10 months to 5.25 years post implant	6(100%) 1 CPVT, 2 HCM, 1 DCM, 1 LQTS, 1 Brugada	Interview
Sleeper et al, 2016 USA	Cross-Sectional Quantitative	355 children in cardiomyopathy registry Median 8.6 years (≥ 5 years) 2.6 years post Dx	355(100%) 149 DCM 129 HCM 77 mixed/other	CHQ Functional Status II
Stefanelli et al, 2002 USA	Retrospective Qualitative	27 patients with ICDs32 ±29 months (range 1-100, median 24 months)	15(56%) 9 LQTS, 6 HCM	Review of medical chart notes
Wojcicka et al, 2008 Poland	Cross-Sectional Quantitative	45 teen/adult patients with ICDs 4.3 ± 2.7 years (range 5 months to 11 years) post implant	36 (80%) 19 HCM, 3 DCM, 5ARVC,6 LQTS, 3 CPVT	Questionnaire developed for the study

P1312 - ECHOCARDIOGRAPHY AND CORONARY ANGIOTOMOGRAPHY AS A NON INVASIVE METHOD FOR DIAGNOSIS OF CORONARY ANEURYSMS IN KAWASAKI DISEASE

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Background: Kawasaki disease (KD) is a systemic vasculitis that mainly occurs in children under 5 years. Coronary artery aneurysms (CAA) occur in 20% of cases untreated with intravenous immunoglobulin (IVIG), and in 5-7% of treated patients, which can lead to ischemic heart disease and death.
Case 1: MG, 2y7m old male, presented a 6-day fever, followed by oropharynx hyperemia, maculopapular rash in trunk and members, bilateral conjunctivitis and hyperemia in hands and feet. The echocardiogram (ECHO) was normal. He was treated with IVIG (2 g/kg) and acetylsalicylic acid (AAS) 100 mg/kg/day until being

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afebrile for 72 h and was discharged with AAS (5 mg/kg/day). A month later, a new ECHO showed a 7mm aneurysm in the right coronary artery (RCA) and 6mm one in the left coronary artery (LCA). A coronary angiotomography (CAG) confirmed the ECHO findings.
Case 2: RDS, 3 year-old male, presented complete symptomatology for KD. On the 7th day of onset disease, he received IVIG (2 g/kg) and AAS (100 mg/kg/day). His fever persisted, so prednisone (2 mg/kg/day) was administered for 14 days. The first ECHO and EKG were normal. He was discharged with AAS (5 mg/kg/day). The follow-up ECHO and CT scan showed a 5mm aneurysm in the RCA and multiple CAA like rosary beads in the LCA. The CAG showed further aneurysms in the RCA and left circumflex artery. Infliximab was administered (5 mg/kg), in an attempt to reduce cardiac sequelae.
Conclusion: KD surpassed rheumatic fever as leading cause of acquired cardiac disease in children of the developed world. Despite the use of IVIG, 10% of patients develop cardiac alterations. CAG complements the evaluation detecting aneurysms, thrombus, and stenosis in the distal regions of the coronaries which the ECHO is unable to identify.

P1315 - OBSERVATIONAL STUDY OF CARDIAC ALTERATIONS IN PATIENTS WITH MUCOPOLYSACCHARIDOSIS

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Background: Mucopolysaccharidosis (MPS) are inherited lysosomal storage disorders caused by the deficiency of enzymes responsible for the degradation of glycosaminoglycans (GAGS). The systemic deposition of GAGS results in progressive organic dysfunction. Cardiovascular involvement is variable but contributes towards the morbidity/ mortality of the patients.
Materials and Methods: A prospective observational study was conducted with 27 MPS patients aged 3-21 years. Two patients were type I, seven type II, three type IV and fifteen type VI. Twenty-three patients underwent enzyme replacement therapy (ERT). Their clinical alterations, electrocardiographs (ECG) and echocardiograms (ECHO) were analyzed. Cardiac findings, time of ERT and severity of valvular lesions were compared between the different types of MPS.
Results: Although ECHO alterations were present in all patients, 74,1% referred dyspnea or palpitations, only 55,6% presented murmurs and 7,4% showed ventricular overload in the ECG. All patients were in sinus rhythm, and 22,2% had incomplete right bundle branch block. Systolic function was overall normal, although 30,8% presented left diastolic dysfunction. Pulmonary hypertension was found in 33,3% and systemic hypertension in 25.9%. One patient had a biologic mitral valve prosthesis. Mitral valve thickening was seen in 26 patients (92.3%), of which 29.1% had no dysfunction, 75% had discrete or moderate regurgitation, and 3.7% had double lesion. The aortic valve was affected in 70.4% of patients (11.1% had double lesion and 59.3% had regurgitation). Mild or moderate tricuspid regurgitation was present in 33.3% and left ventricle hypertrophy in 44% of the patients. The type IV patients manifested less mitral valve lesions ($p = 0{,}035$).
Conclusion: Valvular lesions are frequent in MPS patients particularly in those with type I, II and VI. These patients presented a higher prevalence of mitral and aortic valvulopathy with a predominance of regurgitation.

P1364 - "DOCTOR WHAT WOULD YOU DO IF IT WAS YOUR CHILD – IS THAT THE CORRECT QUESTION TO ANSWER IN PRENATAL CARDIAC COUNSELLING "

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Background: The prenatal diagnosis of a complex cardiac abnormality generates considerable stress for the affected parents. Counselling strategies have been described with limited documentation of their efficacy.
Methods and Materials: Against the background of an accurate fetal diagnosis parents were informed in simple lay terms, aided by diagrams and the attending obstetrician ultrasonologist, of the nature of the abnormality, it's likely consequences and potential interventions. Explanations included the immediate and long term functional outcomes rather than the detailed anatomy. At times multiple visits were required being cognizant of the parental emotional distress and their difficulties to "hear" and assimilate what is said.
Results: Using a self report scale, 70% of mothers of a cohort of 39 affected pregnancies felt that the above approach allowed for a clear or very clear explanation of the cardiac abnormality, 85% felt that the details were just right while 60% rising to 80% following subsequent scans, described their understanding to be good to excellent. These results were especially meaningful when 70% acknowledged being distressed or very distressed at the time. At no stage even if asked, was it necessary for the clinicians to give their own views as if it was their child.
Conclusion: Simple explanations emphasising function and outcome and taking into account the emotional distress of the parents, allowed for a good understanding of the cardiac abnormality and its consequences. Such an approach may have facilitated informed consent if termination was an option. It did away with the clinicians expressing their personal views allowing the parents to reach their own decision taking into account their life experiences, needs and responsibilities.

P1365 - LIPID ABNORMALITIES IN CHILDREN ONE YEAR AFTER KAWASAKI DISEASE

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Background: Kawasaki Disease (KD) patients have abnormal lipid profiles at presentation. Because coronary artery lesions (CAL) develop in 25% of untreated KD patients, lipid profile screening has been recommended by the American Heart Association guideline in KD patients after recovery. High density lipoprotein (HDL) has been shown in adults to be protective of atherosclerosis and coronary artery disease; however, we do not know the role of HDL in KD patients with CAL. In this study, we evaluated the lipid profiles of KD patients 1 year after disease onset and compared lipid profiles of KD patients with and without CAL.
Methods: Patients medical charts were retrospectively reviewed from 2014-2016. CALs were defined as left anterior descending artery and right coronary artery z-scores ≥ 2.5. Lipid profiles were obtained in KD patients (fasting state) 1 year after disease onset. Demographic and biomarkers were compared with HDL $\geq$ 45 mg/dl and $<$ 45 mg/dl and between patients with and without CAL using a student s t-test for normally distributed continuous

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variables, Wilcoxon ranked sum test for non-uniform distributed variables, and $\chi 2$ for categorical variables.
Results: 53% of KD patients had HDL <45 mg/dl. There were no significant differences in clinical and initial laboratory values between the two groups (Table 1) with the exception of hemoglobin ($p < 0.007$) and albumin ($p < 0.01$). There were no significant differences in lipid profiles in KD patients with and without CAL (Figure 1). There were no significant differences between the body mass indexes between the two groups when the lipids were drawn at 1 year after disease onset.
Conclusions: Half of KD patients had HDL <45 mg/dl one year after disease onset. Preventative counseling to KD patients diet and exercise are important in KD patients with abnormal HDL. Future studies are are needed to elucidate the pathophysiology of this lipid abnormality in KD patients.

Table 1. Clinical and Laboratory Characteristics of KD patients at time of diagnosis with low and normal HDL.

	HDL < 45 (n = 39)	HDL ≥ 45 (n = 34)	P-value
Age (years)	4.3 ± 3.1	4.3 ± 3.5	0.9688
age < 1yr	1 (2%)	4 (12%)	0.1040
Gender (M/F)	31/8	24/10	0.3901
fever period	9.8 ± 4.5	9.3 ± 3.4	0.5847
CRP	11.1 ± 7.4	10.5 ± 6.8	0.7752
ESR	59 ± 25	47 ± 24	0.0514
WBC count	16.1 ± 6.7	13.9 ± 5.6	0.1471
Hgb	11.2 ± 2.1	12.0 ± 1.2	**0.0073***
Platelets	350 ± 129	377 ± 118	0.3485
Albumin	3.2 ± 0.7	3.5 ± 0.5	**0.0148***
AST	77 ± 61	66 ± 42	0.4108
ALT	78 ± 50	90 ± 73	0.4546
IVIG + Remicade	7 (16%)	5 (17%)	0.8338

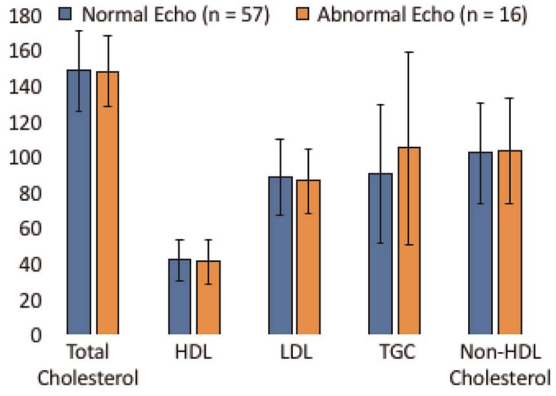

Figure 1.
Lipid profiles between KD patients with and without normal echocardiograms.

P1395 - QUEENSLAND FAMILIAL HYPERCHOLESTEROLAEMIA CLINIC OUTCOMES ANALYSIS

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Background: Consensus guidelines exist for the diagnosis, treatment and management of Familial Hypercholesterolaemia (FH) in childhood. We review the practical outcomes of preventative intervention from a state-wide paediatric FH clinic.
Methods: Prospective assessments included response to treatment, review of side-effects, carotid intima-media thickness (CIMT) and echocardiography at appropriate intervals. Of the 26 clinic patients, 24 had full datasets for lipid profiles including initial response and late follow-up. 14 had CIMT measurements early and at late follow-up.
Results: Presentation was predominantly in asymptomatic children from cascade screening. All patients had phenotypically confirmed FH with elevated low-density lipoprotein (LDL-C) levels (>4.0 mmol/L) and a significant family history. The cohort was 63% male with diagnosis occurring at median age of 12.7 years (range 4.8-16.8). 4 patients had concurrent type 1 diabetes mellitus. Statin treatment was commenced at median age of 13.0 years (range 8.0-16.8) and followed guidelines. At diagnosis mean total cholesterol (TC) was 8.2 mmol/L (CI 0.6) and mean LDL-C was 6.1 mmol/L (CI 0.5). Initial post-therapy levels were TC 5.6 mmol/L (CI 0.3) (=31% reduction, CI 4%) and LDL-C of 3.7 mmol/L (CI 0.4) (=39% reduction, CI 4%). These reductions were maintained at median follow-up of 3.5 years (range 1.1-9.2) with TC 5.7 mmol/L (CI 0.5) (=29% reduction, CI 6%) and LDL-C 3.9 mmol/L (CI 0.5) (=35% reduction, CI 6%). Targeted dose escalation was required in 45% of patients. 8 children changed medications with 6 due to documented mild side-effects, which all resolved. During follow-up Left-CIMT (n=14) was reduced from 0.52mm (CI 0.04) to 0.48mm (CI 0.02). (All reported results $p < 0.05$).
Conclusion: We demonstrated that early intervention has the potential to improve lipid levels and CIMT, both of which are associated with cumulative lifetime risk-reduction. A large proportion of unrecognised and untreated paediatric patients must exist based on known incidence of FH of >1:500.

P1415 - CHARACTERIZATION OF PULMONARY HYPERTENSION IN CHILDREN IN ALTITUDE (2.640 METERS OVER SEA LEVEL) IN LAST YEAR

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Introduction: The effect of altitude is significant over 2.500 MOSL. Bogotá, where we made the study, is at 2.640 MOSL.
Objective: To show the effect of Hypobaric Hypoxia on PH in Children.
Patients: We include only new patients studied the last 12 months. All had Clinical evaluation, BNP, prolonged Hyperoxia Test and Catheterism, except one patient that didn´t have catheterism because he died before the procedure. Necropsy study showed Idiopathic Pulmonary Hypertension with severe Pulmonary Vascular Disease. In the Cath. Lav. all patients had Vascular Reactivity Test with NO and Oxygen. Living at low altitude is recommended.
Results: We had 20 new patients; 16 females (80%); 8 were 3 years old or less (40%); 12 had IPH (60%), 4 CHD (3 developed PH after surgery); 2 CPD, 1 HAPE and 1 group 5. Majority of patients had severe PH: MPP >60 mm Hg; PR >20WU.

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Discusion: Altitude gives special characteristics to PH by hypobaric hypoxia, affecting epidemiology, pathogenesis, diagnostic approach and treatment. Epidemiology: PH is frequent, begins and could be severe at an early age, and IPH is the main cause. Pathogenesis: Hyperreactivity of Pulmonary Vasculature is important and remodeling could be severe very early. Diagnosis and treatment: Oxygen plays important role.
Conclusion: At altitude, Hypobaric Hypoxia gives special characterstics to PH. Because Hypobaric Hypoxia influences not only other pathologies of group 3 but also pathologies of groups 1 and 5 of Nice Classification, maybe PH of altitude deserve to be considered as a subgroup of group 3.

P1421 - EARLY DETECTION OF SEVERE PULMONARY HYPERTENSION IN CHLDREN AT ALTITUDE 2.640 METERS OVER SEA LEVEL

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New advances in knowledge of PH, have allowed longer and better quality of life of patients. It is important early detection for avoid the remodeling of pulmonary vasculature.
Objective: To show a serie of cases with severe PH in children diagnosed in early childhood in Bogota, Colombia (2.640 MOSL), analyzing the outcome and the answer to early diagnosis.
Material and Method: We analyzed clinical and laboratory records of children with severe PH. Diagnosis was made considering clinical findings, EKG, Thorax X Ray, echocardiogram and catheterism. Oxygen and vasodilators were used.
Results: 31 patients had severe PH: media PSP: 102 mm Hg. (68-170). Pathologies: IPH: 23; ES: 2; Talasemia: 1; With CHD: 5; Age at diagnosis: Media: 3.85 years (0.4-12 Years). Time of follow up: 5.3 years; (0.5-33 years). One child (22 months old at diagnosis), had a BMPR2 mutation. Today after 14 years of Follow up, is asymptomatic. 8 patients died, 10 stable, 9 very well, 2 asymptomatic and 2 bad. 18 went to live at low altitude; 13 are well or stable.
Discussion: Initially PH is a "silent" disease, but can be detected early, considering clinical aspects. In altitude, the therapeutic approach is different; it is important to live at low altitude. The early detection of PH correlate with better outcome although by hypobaric hypoxia, PH could be agressive and 8 patients died in spite of live at low altitude
Conclusion: PH is a disease with poor prognosis and early mortality. There isn't cure; with early diagnosis we can get better prognosis, avoiding pulmonary vascular disease. In 31 children we have got a mean follow up of 6.3 years (0.5-33) including a patient with a BMPR2 mutation (initial PSP: 153 mm Hg) with Follow up of 14 years. The patient is asymptomatic with PSP of 48-50 mm Hg.

P1476 - PULMONARY ARTERIAL HYPERTENSION AS A LATE COMPLICATION OF CONGENITAL HEART DISEASE

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Background: Despite advances in diagnosis and treatment of congenital heart disease (CHD), there are many patients with undiagnosed or un-operated CHD in the developing countries, mainly because of late referral to specialized centers. Pulmonary arterial hypertension (PAH) associated with CHD (PAH-CHD) is usually the result of a large systemic-to-pulmonary shunt and it is a heterogeneous condition, depending on the type of the underlying defect and previous repair strategies. The presence of PAH increases morbidity and reduces survival in patients with CHD. The prevalence of PAH-CHD has been estimated to range between 1.6 and 12.5 cases per million adults. Despite of recent evidence of the benefits of PAH-specific therapy in the PAH-CHD population, the mortality rates remain relatively high.
Objectives: determinate the epidemiologic characteristics and outcomes of patients with diagnosis of HAP in a regional referral center in Brazil.
Methods: all patients with PAH-CHD were included. Data about gender, age, underlying diagnosis, previous cardiac surgery, medications in use and mortality were recorded.
Results: from a total of 113 patients in the registry, 70 were female (61,9%). Average age of these patients was 24,13 ± 14,40 years (range from 4 to 76 years) and 18,8% had Down Syndrome diagnosis. The average time of follow up was 6,12 ± 4,09 years. The most frequent underlying defects were ventricular septal defects (20,4%), complete atrio-ventricular septal defect (20,4%) and atrial septal defects (15,9%). Thirty two patients (28,3%) have undergone previous surgical procedures. The majority of patients were using sildenafila (89,1%), furosemide (67,3%) and digoxine (61,9%). Only 10,6% of patients were receiving treatment with bosentana. The overall mortality was 6,2%.
Conclusion: The HAP-CHD is a common complication of late treatment of CHD with high mortality.

P1513 - LIPOPROTEIN (A) DOES NOT ADVERSELY IMPACT VASCULAR HEALTH IN CHILDREN WITH DYSLIPIDEMIA

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Background/Hypothesis: Lipoprotein (a) [Lp(a)] accelerates atherosclerosis and is strongly heritable. Elevation of Lp(a) is an independent risk factor for premature cardiovascular disease in adults, but its role in atherosclerotic progression in children is unclear. We conducted a cross-sectional pilot study to determine whether Lp(a) contributes to altered vasculature in children with dyslipidemia.
Materials & Methods: Children and adolescents aged 9-18 years with dyslipidemia were prospectively recruited from a pediatric preventive cardiology clinic. Each completed a lipid panel and Lp (a) measurement. Participants were separated into either a study group (Lp(a) ≥30 mg/dL) or control group (Lp(a) <30 mg/dL). Pulse wave velocity [PWV], a measure of vascular stiffness, was calculated using a combined oscillometric and tonometric device. Carotid artery intima-media thickness [cIMT] was measured along the far wall of the right and left common carotid arteries using semi-automated edge detection software.
Results: 64 children (32 per group) were recruited: study group (37.5% males, age 15.43 ± 2.35 years) and control group (40.6% males, age 13.42 ± 1.84 years). Number of vascular risk factors was 3.7 ± 1.6 in the study group and 3.4 ± 1.3 in the control group. Family history of premature cardiac death was present in 21.9% of the study group and 15.6% of the control group. Lp(a) was 99.8 ± 55.4 mg/dL (median 93.0 mg/dL) in the study group and

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10.1 ± 4.4 mg/dL (median 10.0 mg/dL) in the control group. Between groups, there was no difference in PWV (study 4.7 ± 0.8 meters/second [median 4.8 meters/second]; control 4.8 ± 1.7 meters/second [median 4.6 meters/second]; p = 0.83) or cIMT (study 0.47 ± 0.05mm [median 0.45mm]; control 0.47 ± 0.04mm [median 0.47mm]; p = 0.61).
Conclusions: Despite the presence of elevated Lp(a) and other atherosclerosis-promoting risk factors in this young, cross-sectional study population, there was no evidence of adverse change in the vasculature when compared to those with normal Lp (a). However, continued follow-up of this high-risk group is recommended in view of their known risk for premature cardiovascular disease.

P1517 - RIGHT ATRIUM ANEURYSM IN A 14 YEAR OLD BOY DIAGNOSED SINCE INTRAUTERINE LIFE

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Background: Right atrial (RA) aneurysm is a rare cardiac abnormality of unknown origin with few cases described in the literature, especially in intrauterine life. This defect is usually diagnosed incidentally and many patients are asymptomatic. Others present arrhythmia, chest pain and shortness of breath. The best approach to asymptomatic patients has been controversial and the natural evolution of these malformations remains uncertain.
Case Report: The authors describe a 14-year-old-boy who was referred with palpitations and shortness of breath during mild efforts. He was using no medication. He had had an intrauterine diagnosis of right atrium aneurysm, with no other malformations and was accompanied in another city. On physical examination he was eupneic and acyanotic. In the thoracic auscultation, the heart sounds were rhythmic with a systolic murmur at tricuspid area with a positive Rivero-Carvallo's sign. Chest radiography disclosed considerable cardiomegaly due to RA dilatation. On electrocardiogram, he presented sinus rhythm with signs of right atrium enlargement. Echocardiogram revealed a normal positioned tricuspid valve, a huge right atrium and right atrium appendage. Right atrium volume was 413 mL. There was moderate to severe tricuspid regurgitation. He underwent surgery with extracorporeal circulation. The right atrium was opened with resection of a large aneurysm and a tricuspid valvuloplasty was performed. Postoperative echocardiogram revealed discrete increased right atrium with a 49 mL volume and mild to moderate tricuspid regurgitation. He was discharged on the tenth postoperative day in good conditions.
Conclusions: For the little experience accumulated with RA aneurysm diagnosed in intrauterine life, the wiser conduct seems to be expectant. Fetal echocardiography plays an important role in diagnosis of congenital cardiopathies and in the event of finding atrial dilatation on obstetric ultrasound examination, fetal echocardiogram should be performed.

P1519 - FOUR NEUROLOGICALLY INTACT SURVIVING PATIENTS AFTER COLLAPSE DUE TO HYPERTROPHIC CARDIOMYOPATHY OUTCOMES OF MANAGEMENT BASED ON HEART SCREENING FOR SCHOOLCHILDREN

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Background: The system of electrocardiogram screening for all schoolchildren was initiated in 1997 in Japan. Since the use of automated external defibrillators (AEDs) by the general public was approved in 2004, many AEDs have been supplied to public schools. More children with hypertrophic cardiomyopathy (HCM) are being successfully resuscitated after collapse owing to the increased availability and accessibility of basic life support systems. Our hospital conducts screening of schoolchildren in Kobe City, and provides emergency and critical care. Four patients with HCM who survived after being resuscitated were sent to our hospital since 2005.
Cases: All the patients were diagnosed with HCM while in primary or junior-high school and they had been restricted from performing exercise. They had no subjective symptoms, and no arrhythmia was detected before the cardiac event. Only case 3 had a family history of sudden death.
Case 1: A 16-years-old boy collapsed while practicing judo during. The teachers immediately requested for a rescue team, and the ambulance crew used an AED.
Case 2: A 14-years-old boy collapsed while running on the school ground. The teachers immediately used an AED.
Case 3: A 21-years-old woman collapsed during her internship work. The attending supervisor immediately used an AED.
Case 4: A13-years-old boy collapsed when he was about to leave school. Bystanders and teachers performed emergency cardiopulmonary resuscitation, and the rescue team used an AED.
After being defibrillated, the patients were sent to our hospital and received low-temperature therapy and implantation of an implantable cardiovascular defibrillator. All of them recovered without any neurological sequelae.
Conclusion: We teach about emergency cardiopulmonary resuscitation in junior high schools and high schools, and spend more educational time at schools with more registered high -risk patients. We hope to reduce the total number of sudden deaths under school supervision through the screening system and activities after diagnosis.

P1542 - BEYOND THE SINUS OF VALSALVA POSITIVE EFFECT OF ANGIOTENSIN II RECEPTOR BLOCKERS ON MITRAL VALVE PROLAPSE IN A RETROSPECTIVE ANALYSIS OF PEDIATRIC PATIENTS WITH MARFAN SYNDROME

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Objectives: Treatment with Angiotensin-II-receptor-blockers (ARB) in patients with Marfan syndrome (MFS) has been well established. Most studies, so far, focused on the the aortic root dilatation. No study has focused on the effect on the regularly appearing mitral valve prolapse (MVP) in MFS. The aim of our study was to analyse the treatment effect of ARB on the degree of MVP in pediatric patients with MFS.
Methods: Since 2008 we investigated 398 patients (11.4 ± 5.5 y) with suspected MFS. 140 patients were diagnosed with MFS. 62 patients showed MVP. We retrospectively analysed database data and 2D-echocardiograms. Because of the high error measuring the actual MVP in millimeter, mitral anular diameter (MAD) was used as a surrogate parameter for MVP in MFS (Pini R, 1989, Circulation). Patients in the treatment group received oral Valsartan, control patients had no treatment. Patients taking beta-blockers or having being operated on the aorta were excluded to clearly indicate group allocation.

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Results: Prevalence of MVP in pediatric Marfan patients was 43,8%. In the control group the z-score of the MAD did not change significantly during the observation period. In patients treated with an ARB the z-score of the MAD was significantly reduced. Detailed data is shown in table 1.
Conclusion: Prevalence of MVP in children with MFS is 40 times higher than in a healthy population (Sattur S, 2010, Exp Clin Cardiol). In our study, treatment with an ARB in pediatric patients with MFS leads to a significant reduction of the MAD as a surrogate parameter for MVP. This first data suggests another positive effect of ARB treatment in MFS beyond the growth restrictive effect on the aorta. Future studies have to analyse greater numbers of patients, the long term effect and develop better imaging methods for the detailed analysis of MVP e.g. MRI.

Table 1. Demographic and mitral valve date before therapy initiation or at first visit (t0) and at last follow up (t1) in patients with MFS without treatment (control) or treated with an angiotensin receptor blocker (ARB).

Demographic data	control group n = 12 (3 female)			ARB group n = 8 (4 female)		p value
Age (years) at treatment initiation /first visit	8,2 ± 4,75			12,3 ± 3,61		ns
Follow up period in years	2,4 ± 1,93			3,4 ± 2,08		ns
Mitral valve prolapse	t0	t1	p value	t0	t1	p value
z-score of mitral anular diameter	2,23	2,04	0,5121	2,59	2,21	0,0336

ARB, angiotensin II receptor blocker

P1557 - ANOMALOUS CORONARY ARTERIES CONNECTED TO PULMONARY ARTERY IN COMBINATION WITH OTHER CONGENITAL HEART DEFECTS AND EXTRACARDIAC ANOMALIES – OVERVIEW OF 98 PATIENTS

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Background: Anomalous coronary arteries connected to the pulmonary artery are rarely associated with other congenital heart defects.
Materials and Methods: From 1974 to 2016, 98 patients with anomalous coronary arteries connected to the pulmonary artery were retrospectively analyzed. Anomalous left coronary artery connected to the pulmonary artery (ALCAPA) was present in 84 patients, anomalous right coronary artery connected to the pulmonary artery (ARCAPA) in 4, anomalous circumflex coronary artery connected to the pulmonary artery (ACXPA) in 6, anomalous left anterior descending coronary artery connected to the pulmonary artery (ALADPA) in 2, and anomalous single coronary artery connected to the pulmonary artery (ASCAPA) in 2 patients. Surgery and angiography protocols as well as medical reports were screened for any type of other cardiac and extracardiac defects and the connecting part of the pulmonary artery.
Results: Abnormal origin of the coronary arteries from the pulmonary arteries is associated in 12% of the patients with other congenital heart defects; the combination of left heart obstructions and ACXPA was observed particularly often. Extracardiac anomalies were found in 2 ALCAPA (Rubinstein-Taybi, pelvic kidney); one ARCAPA had fetal alcohol syndrome and another one VACTERL association; one ACXPA had cystic kidney disease with vertebral anomaly, another ACXPA segmental lung hypoplasia; one ASCAPA had congenital diaphragmatic hernia. In ACXPA patients the anomalous coronary artery connected in 5 of 6 patients from the right pulmonary artery.
Conclusions: Especially in patients with left heart obstructions and heart failure, the possible presence of an abnormal origin of the coronary artery from a pulmonary artery should be taken into account.

Table.

Group	Number of Cases	Combination with other CHD	Associated CHD	Connecting part of pulmonary artery
ALCAPA	84	2	1: CoA + VSD, 2: Left carotic artery from Truncus brachiocephalicus, LPSVC	81 from PA, 1 RPA, 2 LPA
ARCAPA	4	3	1: HLHS, 2: VSD, + ASD, + LPVCS, 3: APW + VSD + LPSVC	3 PA, 1 RPA
ACXPA	6	6	1: CoA + VSD + ASD, 2: CoA + BAV, 3: CoA, 4: HLHS, 5: BAV + AS, 6: CoA + VSD	1 PA, 5 RPA
ALADPA	2	0	-	2 PA
ASCAPA	2	1	1: Shone Complex	1 PA, 1 RPA

P1563 - ASSESSMENT OF QUALITY OF LIFE IN TURKISH CHILDREN WITH CONGENITAL HEART DISEASE

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Objectives: Due to the recently improved survival rates of children and adolescents with congenital heart disease (CHD) greate attention should be focused on outcomes of treatment than survival– such as the patients' quality of life (QoL). The main aim of this study was to assess and compare the self-reported and parent-reported QoL of children with CHD with the healthy controls. We also aimed to determine whether there is any relationship between the QoL and the severity of CHD and patients' sociodemographic characteristics.
Methods: The patient group consisted of 181 children between 5-18 years of age with a diagnosis of CHD and their families. The control group included 200 healthy children between 5-18 years of age and their families. PEDsQL scores and sociodemographic characteristics were compaired between groups.

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Results: The patient group had poorer levels of monthly income and lower parental education levels compared to controls. The highest impact on QoL was noted in the 5-7 years age group in CHD patients and their families, who had lower total score, physical health, psychosocial health, social functioning and school functioning compared to healthy controls and their families. In the 13-18 years age group, there was no difference in QoL scores between the patients and the controls. A regression analysis revealed that the patient's education level, medical treatments and sex were independently associated with QoL.
Conclusions: We found that children with CHD had lower QoL compared to healthy children of the same age and that gap in QoL scores between these two groups closes as child grows up. Higher QoL scores of male patients is an interesting finding and deserves further discussion. We believe that clinicians play a key role in improving QoL in those patients, with a focus on identifying QoL-lowering risk factors and providing the family with the necessary support.

P1566 - SUDDEN CARDIAC DEATH (SCD) IN CHILDREN AND ADOLESCENTS IS SELECTIVE ELECTROCARDIOGRAM (ECG) SCREENING JUSTIFIED

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Background: SCD in otherwise healthy children is a rare but devasting event. Merits of universal ECG screening programs at a specific age versus selective screening in high-risk individuals (young athletes, Metilfenidate treatment and family history of SCD) have been debated. This study compares results in SCD screening in 2 Spanish different young athlete and adolescent cohorts.
Matherial and Methods: Cross-sectional study at Irun's athletes' football team cohort and San Sebastian's High School adolescents' cohort. Screening included a standardized history, physical examination (PE) and 12-lead electrocardiogram (ECG) as recommended by the American Heart Association. Student's t-test was used to compare continuous variables and Chi-square test for categorical changes. Association was assessed by Odds Ratio (OR) estimation and Spearman's rho (r) was used in correlation analyses.
Results: A total of 265 adolescents (mean age: 12,3 ± 0,9 years, 132 males) and 53 athletes (mean age: 14,5 ± 1,7 years; all males) participated in this study. 12 (3,8%) individuals were under Metilfenidate treatment and 26 (8,2%) had family history of SCD. In adolescents group, 4 (1,3%) had abnormal PE and 7 (2,2%) had abnormal ECG (2 Long QT syndrome, 1 Wolff-Parkinson-White pre-excitation, 2 left ventricular hypertrophy and 2 right ventricular hypertrophy). Athletes had longer QRS interval than adolescents regardless Metilfenidate intake or family SCD history confounders (88,1 ms versus 84,2 ms; p = 0,001). No other significant differences were seen in ECG or PE between both groups. Risk of abnormal ECG was 1,9 times (OR confidence interval: 0,2-16,5) higher for SCD family history individuals. There was a good correlation between PE and ECG screening results for cardiac abnormalities (r 0,56; p < 0,001).
Conclusion: No pathological significant differences were found between both groups. This results do not justify selective ECG screening programs. Extensives studies and cost-effectiveness analysis of universal screening programs are required.

P1568 - OUTCOME OF PATIENTS WITH CONNECTIVE TISSUE DISORDERS PRESENTING WITH CARDIOPULMONARY SYMPTOMS AT LESS THAN 6 MONTHS OF AGE

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Background: Children with Marfan syndrome and other aortopathies are rarely symptomatic during early infancy. Early onset of symptoms is thought to confer an adverse prognosis. We sought to evaluate the natural history of patients with a genetically confirmed connective tissue disorder who presented with cardiopulmonary symptoms at <6 months of age.
Methods & Results: Database review was performed and age, genetic mutation and symptoms at diagnoses were recorded. Age at, type of, and indication for surgical interventions were noted. Thirteen patients were identified. Etiology of symptoms included severe mitral insufficiency in 5, concurrent congenital heart defects in 3, airway obstruction in 2 and aneurysmal PDA in 3. Genetic diagnosis included neonatal Marfan syndrome in 5, vascular Ehlers Danlos in 1, Loeys-Dietz in 1, neonatal ACTA2 vasculopathy in 3, familial Marfan syndrome in 1, smad3 aortopathy in 1, and MYH11 aortopathy in 1. Eleven (92%) patients required surgery with an age at initial surgery of 4.5 + /- 4.0 months. Surgical intervention in infancy included tracheostomy in 4, ductus ligation in 2, mitral valve repair in 3, mitral and aortic replacement in 1, pulmonary valvuloplasty in 1, right ventricular outflow patch in 1, and VSD closure in 1. Of the 11 patients who survived beyond infancy, 9 had progressive aortic dilation with 6 (60%) undergoing aortic root replacement at 14.3 + /- 6.7 years. There were 5 (38%) deaths overall secondary to aortic dissection in 1, arrhythmia in 1, heart failure in 1, and multisystem organ dysfunction in 2. There were no childhood cardiac deaths in those who underwent cardiac intervention.
Conclusion: Timely surgical intervention in infants with heart failure secondary to connective tissue disease allows for long term survival.

P1593 - THERMAL MONITORING OF A FOOT SURFACE WITH A NON CONTACT INFRARED THERMOMETER IN PEDIATRIC PATIENTS AFTER CARDIAC CATHETERIZATION

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Introduction: One of the most important complication is arterial thrombosis in lower extremity arteries associated with cardiac catheterization procedure. We sought to evaluate the thermal changes in lower extremity after femoral arterial puncture more quantitative with non-contact infrared thermometer .
Methods: This prospective study consists of 32 patients who underwent femoral arterial puncture for cardiac catheterization. Surface temperature measurements were done in a stable room temperature (22-24 °C), just before ,immediately after, at 1st, 3rd, 6th, 12th and 24th hours of cardiac catheterization procedure with non-contact infrared thermometer . Measurement were done from left or right foot surface in order to catheterization site. Three consequent measurements were detected and the mean of these

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measurements was taken. No signs of arterial thrombosis was detected in any patient.
Results: Mean foot surface temperature decreased significantly ($p = 0.00$) immediately after the procedure. After one hour the procedure, foot surface temperature reached the pre-procedural level and continued to rise. During follow up temperature was detected to rise gradually at 2., 3 and 12. hours after completion of procedure ($p = 0.00$). The last measurement was done after 24 hours and the mean temperature value was declined and closed to the initial pre-procedural value ($p = 0.915$).
Discussion: Our results demonstrate that skin temperature was significantly decrease immediately after the arterial puncture. In the following monitoring it was found that the surface temperature increased gradually and reached the highest value at 12 hours after the procedure. Finally it was found that it returned to the initial value measured before the procedure. This temperature changes suggested that reactive hyperthermia occurs in the affected extremity shortly after the procedure and it returns to normal values after 24 hours.

P1611 - LEOPARD SYNDROME ASSOCIATED WITH HYPERTROPHIC CARDIOMYOPATHY

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Backround: LEOPARD syndrome is a rare genetic disorder that affects multiple organs. Characteristic symptoms include: lentigines, electrocardiographic conduction abnormalities, ocular hypertelorism, pulmonary stenosis, abnormal genitalia, retardation of growth and deafness (sensorineural). Cardiac involvement may have a significant impact on the prognosis.
Case Presentation: We report two cases of LEOPARD syndrome in an 18-year-old boy and a 10-year-old girl. In both cases multiple lentigines, ocular hypertelorism and growth retardation were present together with obstructive hypertrophic cardiomyopathy. Both patients met the criteria for LEOPARD syndrome. The second case was caused by a mutation in the PTPN11 gene. The first patient underwent surgery for a moderate obstruction of the left ventricular outflow tract with uneventful postoperative follow up. The presented cases fulfill the criteria for a typical LEOPARD syndrome with predominant left ventricular outflow tract obstruction.
Conclusions: The diagnosis of hypertophic cardiomyopathy in children and adolescents is important because there is a risk of sudden death. Besides traditional clinical findings, hypertrophic cardiomyopathy with left ventricular outflow track obstruction may be an improtant diagnostic and prognostic parameter in patients with LEOPARD syndrome.

P1614 - ARTERIAL STIFFNESS IN CHILDREN WITH WHITE COAT AND ESSENTIAL HYPERTENSION

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Introduction: The term "arterial stiffness" denotes alterations in the mechanical properties of arteries, which may represent higher cardiovascular risk in middle-aged and older adults. Recently, the question regarding the arterial stiffness in patients with risk factors for the early progression of systemic atherosclerosis in vulnerable adolescent age-period are rare. Thus, the aim of this study was to evaluate the carotid stiffness and early atherosclerotic changes in adolescents suffering from essential and white coat hypertension using noninvasive markers.
Methods: One hundred thirty eight children – 46 (23 boys) children suffering from WCH, 46 (23 boys) patients with essential hypertension (EH) and 46 age/gender-matched healthy controls – were examined under standard conditions. The age-period of all children ranged from 14 to 18 years. Ultrasonography combined with echo-tracking system (Prosound F75 Aloka) on common carotid artery (CCA) was used to analyse of local arterial stiffness. In the study five physiological parameters of the CCA were analysed – mean stiffness index (β), elastic modulus (Ep), arterial compliance (AC), augmentation index (AI) and pulse wave velocity (PWV β).
Results: Statistical analysis revealed significant differences in parameters β, Ep, AC and PWV β between the both hypertensive groups (WCH, EH) and control group (Tab 1). Significant differences were not found in the index AI between EH, WCH and control groups. No significant differences were found between WCH and essential hypertensive groups.
Conclusions: Study revealed significantly increased carotid stiffness in children suffering from white coat and essential hypertension. This finding could help to understand hypertension atherosclerosis interaction. We suggest that further research regarding the potential atherosclerotic changes using the sensitive noninvasive parameters in pediatric hypertension is important. Support: VEGA 1/0087/14, project "Biomedical Center Martin" ITMS code [26220220187] co-financed from EU sources.

Table 1. Parameters of carotid stiffness in children with white coat hypertension (WCH), essential hypertension (EH) and control group (C group).

	WCH n = 46	EH n = 46	C group n = 46
β	6,14 ± 0,9***	6,01 ± 1,01***	4,8 ± 0,9
Ep (kPa)	85,4 ± 14,4***	85,2 ± 16,4***	56,1 ± 9,7
AC (mm^2/kPa)	0,84 ± 0,2***	0,84 ± 0,18***	1,10 ± 0,24
AI	5,06 ± 37^{NS}	6,7 ± 32^{NS}	-6,1 ± 6,7
PWV β (m/s)	5,3 ± 0,5***	5,3 ± 0,6***	4,4 ± 0,4

β, mean stiffness index; Ep, elastic modulus; AC, arterial compliance; AI, augmentation index; PWV β, pulse wave velocity ***, $p < 0,001$ compared to control group.

P1620 - ADOLESCENT HYPERTENSION IS ASSOCIATED WITH DIMINISHED SYMPATHETIC REACTIVITY TO STRESS ASSESSED BY NONLINEAR ANALYSIS OF HEART RATE VARIABILITY

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Background: Excessive sympathetic activity is a well-known mechanism involved in pathophysiology of hypertension and related cardiovascular complications. Importantly, both dysregulation of basal autonomic outputs and the altered reactivity to stress increases the risk of cardiovascular diseases. Therefore, we aimed to assess the complex cardiac sympathetic regulation under stress conditions using novel method of non-linear heart rate variability (HRV) analysis – symbolic dynamics - in adolescents with newly diagnosed essential and white-coat hypertension.
Materials and Methods: Thirty adolescent boys with newly diagnosed essential hypertension (16.9 ± 0.2 years, body mass index (BMI) 22.9 ± 0.4 kg/m2), thirty patients with newly diagnosed white-coat hypertension (16.8 ± 0.3 years, BMI 22.6 ± 0.5 kg/m2) and thirty healthy controls (16.2 ± 0.4 years, BMI 22.1 ± 0.4 kg/m2) were examined. Electrocardiogram was continuously recorded in supine position and during active orthostatic test (phase duration6 minutes). Sympathetic cardiac regulation was assessed using nonlinear analysis of HRV complexity - symbolic dynamics index 0 V% (zero variation of HRV).
Results: The beta-adrenergic cardiac activity indexed by 0 V% was significantly increased during supine position in both white-coat and essential hypertension groups compared to control group ($p < 0.05$ for both). The reactivity (percentual increase) of sympathetic index 0 V% to orthostatic test was significantly lower in white-coat and essential hypertension compared to control group ($p < 0.05$ both). No significant between-groups differences were found in absolute values of 0 V% during orthostatic test.
Conclusions: The novel sympathetic marker- symbolic dynamics HRV index 0 V% - was sensitive to assess greater sympathetic cardiac activity at rest, and lower reactivity to orthostatic stressin both white-coat and essential hypertension. Our findings could help to elucidate the role of diminished physiological reactivity to stress in the pathophysiology of hypertension and possible negative health outcomes.
Support: This work was supported by grant VEGA 1/0087/14, APVV-15-0075, andthe project „Biomedical Center Martin"ITMS code: 26220220187, co-financed from EU sources.

P1625 - THE CLINICAL FEASIBILITY OF PRE OPERATIVE NEUROLOGICAL AND NEUROMOTOR ASSESSMENT IN INFANTS UNDERGOING OPEN HEART SURGERY

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Introduction: Infants with congenital heart disease (CHD) who undergo open-heart surgery are at high risk for long-term neurodevelopmental delay. Neurodevelopmental impairments can lead to an increased burden on families, health care systems and educational facilities. Conducting pre-operative assessment allows for early identification of at risk infants and implementation of early intervention. Challenges undertaking pre-operative neurodevelopmental assessments have been reported, limiting the extent to which the impact of surgery can be estimated. This study aims to investigate factors impacting the clinical feasibility of pre-operative assessment in infants undergoing open-heart surgery before four months of age.
Methods: Infants with CHD who underwent open-heart surgery before four months of age participated in this prospective cohort study. The Test of Infant Motor Performance (TIMP) and Prechtl's Assessment of General Movements (GMs) were undertaken pre- and post-operatively. When assessments could not be undertaken, reasons were ascribed to either infant or environmental circumstances. Demographic data and Aristotle scores, which measure complexity of surgical procedures, were compared between groups of infants who did or did not undergo assessment. Associations were explored using binary logistic regression.
Results: Sixty infants were recruited to the study. Median gestational age was 38.78 weeks (IQR 36.93–39.72). Of these infants, 37 (62%) were unable to undergo preoperative assessment due to infant (40%) and environmental (22%) related factors. For every point increase in the Aristotle Patient Adjusted Complexity (APAC) score, the infants likelihood of being able to undergo pre-operative assessment decreased by 35% (OR: 0.35; 95% CI: 1.03-1.77, $p = 0.03$).
Discussion: Over half the participants were unable to undergo pre-operative assessment, primarily due to infant related medical instability. This study highlights the fragility of this cohort and the limitations associated with administering neurodevelopmental assessments pre-operatively. Further research investigating the APAC score as a potential indicator of children requiring developmental surveillance is warranted.

P1645 - WEIGHT TRAJECTORIES ARE ASSOCIATED WITH EXERCISE CAPACITY AMONG CHILDREN WITH COMPLEX CONGENITAL HEART DEFECTS DOES LIMITED GROWTH INDICATE LIMITED ENERGY FOR EXERCISE

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Background/Hypothesis: Children with congenital heart defects (CHD) are often short and lightweight relative to healthy peers, requiring supplementary feeding to meet energy demands. It was hypothesized that CHD patients with limited growth would also have decreased energy for exercise.
Materials and Methods: A retrospective review of Bruce protocol treadmill exercise test results identified CHD patients 8 to 14 years of age. Weight at each clinic visit was extracted from medical charts. Latent class analysis grouped children by growth trajectory: WC 1 = normal weight, WC 2 = underweight, WC 3 = overweight. Extrapolation of energy consumption (ml O2/kg/min) as a function of heart rate standardized energy consumption at a heart rate of 170 bpm for each child. ANOVA examined standardized exercise capacity by weight class, age, and sex.
Results: The growth trajectories of 51 children with CHD (39% female) were classified as normal (n = 10), underweight (n = 19) or over weight (n = 22). Exercise capacity differed significantly by growth trajectory ($p = .003$), but not age ($p = .10$) or sex ($p = .84$). Normal weight children had the highest exercise capacity (estimated marginal mean ± SE: 42.8 + /-2.0 ml O2/kg/min). Overweight children had significantly lower exercise capacity (37.8 + /-1.5, $p = .04$), but underweight children had the lowest exercise capacity (33.8 + /-1.5, $p = .003$), a decrease of 21%.
Conclusion: Children with CHD whose growth trajectory is underweight have much lower capacity for exercise than CHD

patients whose growth is normal or overweight. Insufficient energy for normal growth may increase the risk of sedentary lifestyles, and the associated negative health outcomes. Future research should evaluate whether underweight growth trajectories are associated with decreased daily physical activity and could be used to identify children in need of kinesiology referral.

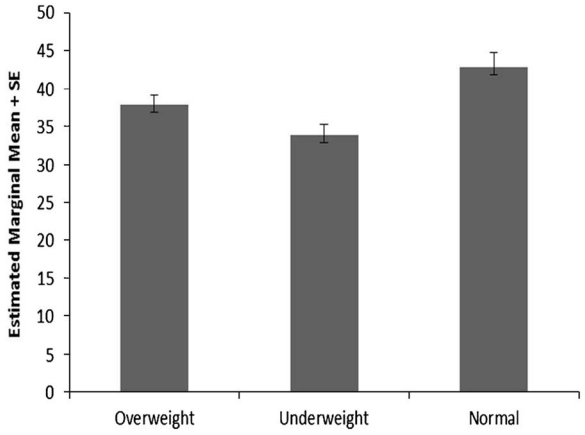

Figure 1.

P1648 - ANATOMICAL AND FUNCTIONAL ASSESSMENT OF CORONARY ARTERIES AFTER ARTERIAL SWITCH OPERATION

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Background: Arterial switch operation (ASO) is the treatment of choice for transposition of great arteries (TGA). However, late complications, such as aortic regurgitation, pulmonar supravalvar stenosis and coronary obstruction may arise in up to 30% of patients. The true incidence and clinical significance of coronary obstruction in these patients is unknown.
Hypothesis: This research aims to correlate anatomical alterations in patients submitted to ASO with rest echocardiography and cardiorespiratory test.
Methods: Sixty-one patients aged five years or above (9.7 ± 3.1 years) were submitted to cardiopulmonary test (treadmill protocol), electrocardiography, rest echocardiography, and coronary artery computerized tomography (CT).
Results: Only two patients (3.3%) presented with severe obstruction of one coronary artery (1 main left; 1 right coronary) although cardiopulmonary test, ECG (both at rest and at effort), and rest echocardiography were normal. Both patients were asymptomatic.
Conclusions: Coronary artery obstruction does not correlate with cardiopulmonary test or rest echocardiography in this population. Coronary artery CT scan is a sensitive and safe method to diagnose coronary artery obstruction in children after ASO.

P1684 - RELATION BETWEEN HEART RATE VARIABILITY AND EXERCISE HEART RATE IN CHILDREN WITH CONGENITAL HEART DIESASE AND HEALTHY CONTROLS

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Background & Hypothesis: Children with congenital heart disease (CHD) display resting autonomic dysfunction as shown by a reduction in heart rate variability (HRV) compared to healthy control children. Children with CHD also display exercise intolerance compared to healthy children that may be due, in part, to a lower exercise heart rate response. We tested the hypothesis that the power spectral HRV ratio would be negatively correlated with 6-min walk test exercise heart rate in children with CHD and healthy control children.
Materials & Methods: Twenty-two children with CHD (11 ± 2 years; females = 9) and 21 healthy controls (11 ± 3 years; females = 11) were studied. CHD diagnoses included simple and complex lesions. HRV was determined following 10 min supine rest using a 5 min surface ECG recorded epoch. 6-min walk test heart rate by telemetry was determined as the average exercise value. Analyses were completed using unpaired t-tests and correlation-regression with $P < 0.05$ being significant. Data are mean ± SD.
Results: 6-min walk test distance was lower (513 ± 75 versus 599 ± 81 m; $P < 0.001$) and 6-min walk test heart rate was lower (122 ± 15 versus 139 ± 18 beats/min; $P = 0.001$) in children with CHD versus healthy controls, respectively. Power spectral HRV analyses revealed no difference in low frequency (LF) power (30 ± 16 vs. 25 ± 16%; $P > 0.05$), but significant differences in high frequency (HF) power (34 ± 19 vs. 49 ± 14%) and the LF/HF ratio (1.4 ± 1.5 vs. 0.6 ± 0.3%) in children with CHD versus healthy controls, respectively (all $P < 0.05$). The LF/HF ratio was significantly correlated with 6-min walk test heart rate for the study group ($r = -0.328$, $P = 0.032$).
Conclusions: Resting autonomic function in children with CHD and healthy controls significantly predicts exercise heart rate. Autonomic heart rate control may be important in the pathophysiology of exercise intolerance in children with CHD.

P1701 - ATHEROSCLEROSIS RISK AND CAROTID INTIMA MEDIA THICKNESS AFTER KAWASAKI DISEASE IN MEXICAN CHILDREN

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Background: Kawasaki disease (KD) is an febrile illness characterized by systemic vasculitis of unknown etiology. Recent studies have shown that endothelial dysfunction persists and may progress to premature atherosclerosis. Carotid intima-media thickness (cIMT) is a well-established marker for atherosclerosis in both pediatric an adult patients.
Objective: To assess whether patients after Kawasaki disease (KD) have increased risk factors and abnormalities suggestive of premature atherosclerosis by measuring the cIMT compared with healthy control subjects
Material and Methods: 153 patients with KD aged 101.05 ± 42.78 months (67.64 ± 39.89 after the acute episode) and 162

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age-matched healthy control subjects were examined for family, medical and dietary history, serum markers of atherosclerotic risk and inflammation and carotid intimal-medial thickness (CIMT) with vascular ultrasound scanning.
Results: Patients and control subjects were similar in age, gender, family and dietary history, body mass index and blood pressure. The levels of total cholesterol (159.37 ± 28.3 vs 147.4 ± 18.3), LDL cholesterol (95.33 ± 25.75 vs 89.63 ± 10.52), and HDL cholesterol (49.88 ± 13.37 vs 39.70 ± 9.28) were slightly higher in patients after KD with no statistical significance. The cIMT was also higher in the KD group (0.48 ± 0.1 vs 0.45 ± 0.15) We did found higher levels in the lipid profile and in the cIMT in children after KD with or without regression of coronary aneurysms compared with children with KD without coronary aneurysms.
Conclusions: There is no clear evidence of increased atherosclerosis in Mexican children with KD, but there is evidence of an altered lipid profile and in the cIMT in patients with KD with coronary lesions compared with children with KD without coronary lesions the warrant further study.

P1710 - DOES ECHOCARDIOGRAPHY HAVE A ROLE IN THE MANAGEMENT OF CONGENITAL HEART DISEASE

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Background: Innocent murmurs are not uncommon in well children. While echocardiography is able to diagnose almost all congenital cardiac abnormalities, previous studies have shown that it may be unnecessary when the clinical diagnosis of an innocent murmur is made by an experienced cardiologist. This study was undertaken to determine if echocardiography had a role in the diagnosis of innocent murmurs in childhood and whether it reduced parental anxiety.
Materials and Methods: Parents of children referred for evaluation of a murmur completed State- Trait Anxiety Inventory questionnaires after the cardiologist's clinical diagnosis of an innocent murmur. The questionnaires were repeated after echocardiography to assess if there was a change in the parental understanding regarding their child's murmur, and to review their level of anxiety. The paediatric cardiologist also completed a questionnaire before echocardiography to determine the reasons for ordering investigation(s), level of confidence regarding his clinical diagnosis, and whether the diagnosis was correct.
Results: 50 mothers have been recruited. 30 of them completed the questionnaires. All 50 echocardiograms were normal except for three. The abnormal changes include congenitally corrected transposition of great arteries and another partial anomalous left upper lobe pulmonary vein both of whom had innocent murmurs. The third revealed a small muscular ventricular septal defect (VSD). Parental and the referring doctor's expectations were found to be the leading reason compelling the specialist to order further investigations. There was a reduction in the anxiety level after completion of the echocardiogram. Enhancement in parental understanding of the diagnosis was also demonstrated.
Conclusions: While the sample size to date is small, the trend shown in this study suggests that echocardiography may be useful not only to clarify the diagnosis but also to supplement parental education and reduce parental anxiety. It also unexpectedly detected three abnormal findings that would otherwise have been missed without echocardiography.

P1718 - PATIENT AND REFERRAL CHARACTERISTICS FOR CLINICAL CARDIOPULMONARY EXERCISE TESTING IN A PEDIATRIC HOSPITAL IN SINGAPORE

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Background: Cardiopulmonary exercise testing (CPET) is a well-established procedure for the evaluation of functional capacity in pediatric clinical settings. It is a tool that can provide useful information to assist pediatricians with diagnosis and management of common pediatric medical conditions. KK Women's and Children's hospital started CPET service in 2009. Till date, there has been limited information on how the CPET service has been utilized in the hospital. The aim of this paper is to describe the characteristics of patients referred for cardiopulmonary exercise testing.
Methods: A retrospective review of patients referred for cardiopulmonary exercise testing, from the year 2009, were conducted.
Results: A total of 37 patients (29 males, 8 females) were referred for CPET since year 2009 and their average age, height and weight were 14.6 ± 2.3 years, 163.5 ± 10.9 cm and 59.5 ± 14.9 kg respectively. Majority of the patients have an underlying congenital heart condition such as tetralogy of fallot (TOF), and dilated cardiomyopathy. Common referral reasons were to evaluate functional capacity, reasons for chest pain, shortness of breath and presyncope. Majority of the test were conducted on treadmill (n = 31), cycle (n = 5), and one patient did not complete the test. Average test duration were 11 mins and 13 secs (range 6 to 16 mins). A summary of the results from the CPET is shown in Table 1.
Conclusion: The aim of this study was to describe the characteristics of patients referred for CPET. Majority of the patients were referred from cardiology service, with majority having congenital heart conditions and a varied range of referral indications.

Table 1. Physiological results from cardiopulmonary exercise test.

Variables	Results
Peak VO^2 (L/min)	2.12 ± 0.8
Peak VO^2 (ml/kg/min)	39.20 ± 10.8
Peak VCO^2 (L/min)	2.51 ± 1.0
RER	1.19 ± 0.1
VE (L/min)	75.95 ± 28.9
VE/VO^2(slope)	39.49 ± 11.1
VE/VCO^2 (slope)	30.88 ± 4.7
Peak HR (bpm)	162 ± 30

P1728 - CHARACTERISTICS OF CARDIOVASCULAR DISEASE IN CHILDREN EXAMINED IN THE CARDIOLOGY OUTPATIENT SERVICE IN A TERRITORIAL HOSPITAL FROM ROMANIA PROSPECTIVE CLINICAL TRIAL

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Introduction: Pediatric cardiology is a relatively new specialty in Romania and there are a small number of pediatric cardiologists, therefore children's with heart disease are being followed up by cardiologists. PURPOSE: The aim of this clinical study was to describe the characteristics of cardiovascular disease in children who were examined in the cardiology ambulatory between 2012 and 2015.
Methods: A number of 422 children from an outpatient cardiology service were enrolled in a prospective study. We recorded demographic characteristics and pathological findings in echocardiography, EKG and Holter EKG evaluation.
Results: Our population consisted of 220 males (52%) and 202 females (48%), with ages between 1 month and 17 years, mean age 5.69. We divided them in age categories and we obtained a number of 53 infants, 118 toddlers, 87 preschool children, 107 school children and 57 in the early adolescence. The most common cardiovascular entities were: mitral valve prolapse in 38 patients(8.9%), patent foramen ovale in 25 patients(5.9%), ventricular septal defect in 28 patients(6.6%),moderate mitral insufficiency in 7 patients(1.65%), moderate aortic valve insufficiency in 2 patients(0.47%), aortic bicuspidy in 4 patients(0.94%), persistent arterial duct in 7 patients(1.65%), atrial septal defect in 14 patients, Fallot's Tetralogy in 2 patients and atrioventricular canal in 2 patients. There was 1 patient with situs inversus and 1 patient with interatrial septum aneurysm. The EKG findings revealed 1 patient with ventricular extrasystole, 1 patient with WPW syndrome and 4 with RBBB.
Conclusions: A complete overview of cardiovascular diseases in children from Maramures County isn't possible because access to pediatric cardiology care is still limited. VSD, ASD and TOF were the most common congenital heart desease in our survey. This study highlights that the development of pediatric cardiology in the region is mandatory.

P1742 - EKG KEY FOR LATE ALCAPA DIGNOSIS

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13 years old presented to clinic of unspecified symptoms easily tiredness , her exam revealed soft systolic murmur at the apex EKG showed q wave in aVL and posterior leads when suspicion of ALCAPA appeared ECHOCARDIOGRAM done with report of mild mitral regurgitation LVd at upper normal Coronary angiogram done shoed evidance of ALCAPA After that patient transferred to a cardiac center for the surgical which done with no cmplication By concentrated reading of the EKG , the diagnosis made and prevent patient from sudden cardiac arrest

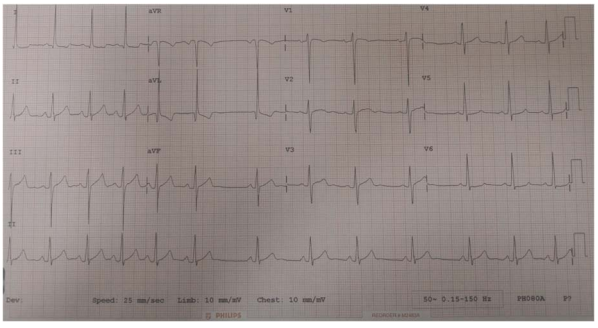

Figure 1.

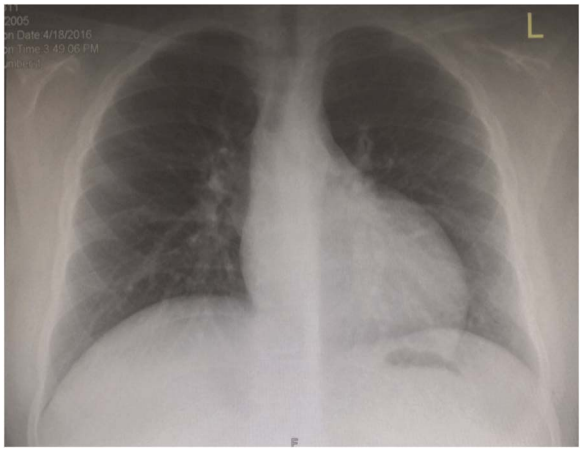

Figure 2.

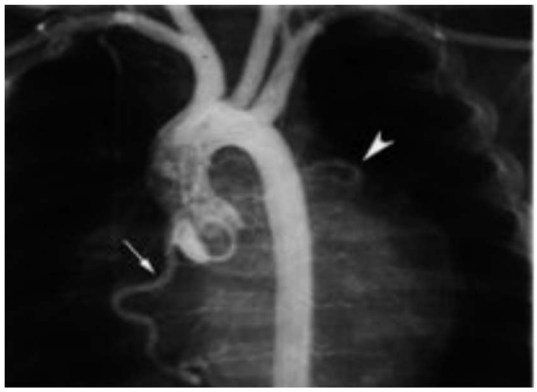

Figure 3.

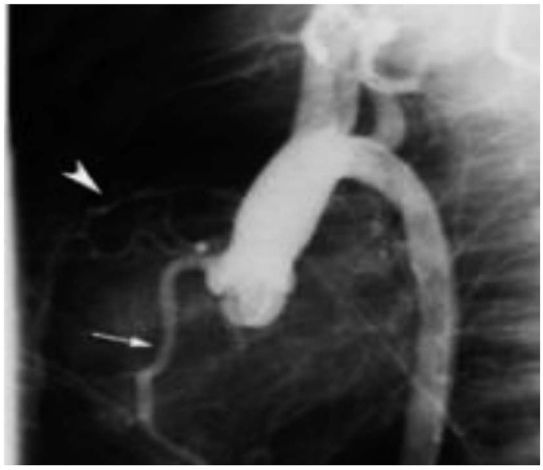

Figure 4.

P1748 - VALUE OF THE OXYGEN DEFICIT AND THE EXCESS POST EXERCISE OXYGEN CONSUMPTION TO EVALUATE EXERCISE PERFORMANCE IN CHILDREN AND ADOLESCENTS WITH CONGENITAL HEART DISEASE

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Background/Hypothesis: during the first minutes of a physical activity there is a delay in the respiratory and circulatory systems to provide oxygen to the active muscles (Oxygen Deficit;OD) and to reach an steady-state. At the end of the exercise there is a high oxygen consumption (Excess Post-exercise Oxygen Consumption;EPOC) to restore the disturbance of homeostasis produced. We hypothesize that physical training in children and adolescents with congenital heart disease (CHD) has a positive impact on OD and EPOC.
Objetive: To determine the usefulness of OD and EPOC measurement to evaluate physical performance in children and adolescents with CHD.
Material and Methods: We analyzed the cardiopulmonary tests (CPT) of our database of the pediatric cardiovascular rehabilitation program, of 25 patients (P) with CHD (15 male – 10 female), aged 5 to 19 years (mean 13). CPT was performed before and after the program (24 session/12 weeks of aerobic exercises) and then the Oxygen Consumption (VO2), the production of carbon dioxide (VCO2) and Minute Ventilation (VE) were analyzed to determine the OD and EPOC in each of the CPT. Lasting of the exercise testing (minutes), heart rate at rest (bpm) and VO2 peak (ml/min/Kg), were used to determine the improvement in aerobic capacity after the program. Matched variables were analyzed with McNemar's test.
Results: the magnitude of OD and EPOC post program was 21% and 24% less than the pre program respectively ($p < 0.05$), and the time elapsed in both was 25% and 30% less than the pre program respectively ($p < 0.05$). Lasting of the exercise testing and VO2 peak increased 70% and 24% respectively ($p < 0.05$), and heart rate at rest decreased 16% ($p < 0.05$) post program.
Conclusions: OD and EPOC can be important surrogates for other more complex measurements and have value to evaluate exercise performance in children and adolescents with CHD.

P1761 - TRANSITION OF PEDIATRIC PATIENTS WITH MARFAN SYNDROME TO ADULT CARE AN IMPORTANT FACTOR TO PREVENT EXACERBATION

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Objectives: Marfan-Syndrome (MFS) is a genetic disorder with a variable clinical presentation. One of the main causes of mortality is the transformation of the cardiovascular system. Aim of this study is the validation of the clinical presentation and status of patients with MFS at date of transition to adult care.
Methods: Since 2008 we investigated 381 patients (11.4 ± 5.5y) with suspected MFS, 51 patients of that group have a diagnosed MFS and are about or have already transited to adult care (age 16-26y; middle age 19.6 ± 2.6y; male:female 28:23). We included most serious manifestations: dilatation of sinus valsalvae (SV), atrioventricular valve prolapse (AVVP), ectopia lentis (EL), skeletal abnormalities (SK, minimum 3 positive signs of revised ghent nosology) ($n = 51$). We also included the incidence of dural ectasia (DE) ($n = 45$), as well as medical and surgical treatment.
Results: All patients show a high percentage of clinical manifestations (SV66.7%, AVVP56.9%, EL13.7%, DE47.1%, SK74.5%). There is a noticeable difference in the level of percentage of patients <18y and >18y (SV75 and 64%, AVVP50/59%, EL8/15%. DE33/51%, SK83/72%). In both groups cardiovascular and skeletal system are more likely affected than lenses and dura. 55% recieve beta blockers and/or AT1 antagonists. 10% have already undergone valve-sparing aortic root replacement. 49% of the >18y old MFS patients are still in pediatric care, 41% have already transited. Amount of lost follow-up examinations was 10% (5/51) only.
Conclusions: It became clear that disease specific pathologies are very dynamic during the period of transition to adult care. The majority of our patients with MFS between 16-26 years of age already show a high percentage of abnormalities of the cardiovascular and skeletal system, irrespective of whether they are over or under 18 years of age. This proves once again how important it is to closely monitor the transition of these patients during this critical episode.

P1762 - CORONARY ARTERY ANEURYSM REGRESSION AFTER KAWASAKI DISEASE IN A MEXICAN POPULATION

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Background: Coronary artery aneurysms (CAA) in Kawasaki disease (KD) developed in 30% of untreated patients and in 3% of patients treated with intravenous gamma globulin. Regression of the CAA has been reported to occur in about 50% of the cases.
Objective: To evaluate regression rate of CAA in Mexican children with KD and evaluate factors associated with regression.
Material and Methods: We evaluated all KD patients with CAA at the Instituto Nacional de Pediatria in Mexico City, Mexico from August 1995 to December 31st, 2015. Regression rate and regression factors were analyzed.
Results: A total of 472 patients with KD were diagnosed at our Hospital during the study period. 154 of them developed CAA (32%). Only 132 of them have been evaluated at follow-up. CAA regression occurred in 91 patients (68.93%). Regression occurred at 11.2 ± 17.35 months after the acute event of KD. Age less than 12 months of age ($p < 0.010$), diagnosis before 10 days after the onset of fever ($p < 0.000$) and small coronary aneurysms based in z-score classification ($p < 0.001$) were the only factors associated with regression of the CAA.
Conclusion: In our study, regression occurs in a slightly higher rate than reported in the literature. Smaller age at diagnosis of KD and small CAA were favorable factors associated with regression.

P1785 - GENETIC VARIABILITY AS A POSSIBLE EXPLANATION OF THE WIDE ANATOMICAL SPECTRUM ASSOCIATED TO OBSTRUCTIVE LEFT HEART DISEASES

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Background: Obstructive left heart diseases (OLHD) encompass a spectrum of various degrees of underdevelopment of left side

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structures. The genetic background of this spectrum has not yet completely known. We aim to identify significant genetic variants and find a possible correlation with different anatomic features in patients with OLHD.
Matherial and Methods: The genomic DNA of 96 patients with OLHD and available parents were studied by TruSeq Custom Amplicon and MiSeq platform (Illumina, San Diego,CA) targeting all the exonic regions of 6 genes associated to the development of valves and ventricles: NOTCH1, LRP6, KIA00182, ZNF236, ZNF423, JARID2.The Exome Sequencing Project (ESP) database was used as normal population. To evaluate a correlation with different phenotypes patients,according to universally recognized echocardiographic criteria ,were divided in hypoplastic left heart syndrome (HLHS), hypoplastic left heart complex (HLHC) and aortic valve stenosis (AVS) group.
Results: 66 single nucleotide variants (SNV) were identified in OHLD. The number of SNVs in OLHD was greater than in ESP population and JARID2 was the more frequently mutated gene (21,9% OLHD versus 1,1% ESP; $p < 0{,}001$). The analysis of variants with a minor allele frequency (MAF) <1% indicated that all 6 genes in OLHD are characterized by very rare variants not reported in any databases (MAF = 0). Moreover the low MAF of these SNVs in ESP population (range 0.029 to 0.043) confirmed that these mutational events are not well tollerated in normal population. A minor number of SNVs was found in patients with hypoplastic left heart valves (HLHC) compared to atresic or stenotic valves (HLHS and AVS) $p = 0{,}023$.
Conclusion: OLHD spectrum could be caused by a cumulative effect of rare low-penetrance alleles superesposed on an unfavorable environment (multi-factorial inheritance model). Preliminary data would suggest a correlation between genetic and anatomical heterogeneity. Further analysis will investigate prognostic and therapeutic implications of our results.

P1792 - LONG TERM FOLLOW UP OF GIANT CORONARY ARTERY ANEURYSMS AFTER KAWASAKI DISEASE IN A MEXICAN POPULATION

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Background: Giant coronary artery aneurysms (GCAA) are a rare but severe complication after Kawasaki disease. The clinical course in these patients often is complicated with ischemic changes either in the acute or chronic phase of the disease.
Objective: Evaluate the long-term follow-up of giant coronary artery aneurysms after Kawasaki disease in a Third-Level Pediatric Hospital in Mexico City, Mexico.
Material and Methods: From the Institutional database of KD, we analyzed all patients who developed GCAA (z-score >10) from August 1995 to December 2016. Information on patient demographics, catheter and surgical interventions, and most recent status was collected from medical records.
Results: 38 patients developed GCAA in the acute phase of KD. Median age at diagnosis was 12.5 months (2 to 66 months). One patient died at diagnosis and one patient was lost to follow-up. Therefore, we analyzed 36 patients. Median time from diagnosis to the most recent medical evaluation was 92 months (5 to 192 months). At follow-up 14 patients (38.8%) developed regression of the coronary aneurysms, median time to regression was 26.5 months. 18 patients (50%) persist with coronary aneurysms, (2 with GCAA). Seven patients developed coronary stenosis in one or more coronary arteries, but only 3 developed myocardial ischemia. CABPG was performed in these patients with a median time from the onset of KD to time of surgery of 105 months (12 to 156 months). One patient developed dilated cardiomyopathy and died 1 year after coronary surgery. The overall survival rate in our series is 94.6%
Conclusions: Despite being a small series, the long-term survival of patients with KD complicated by giant coronary aneurysms in our center is relatively good. Further research, should focus on long-term complications and the indications of coronary interventions.

P1793 - KNOWLEDGE BASE OF ADOLESCENTS WITH CONGENITAL HEART DISEASE

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Introduction: Most congenital heart disease [CHD] is diagnosed in early childhood with parents as decision-makers and recipients of information. Adolescents gradually assume this role. We performed an observational study to evaluate adolescent and parental recognition of their CHD.
Methods: Established clinic patients, >11 y.o. and parents were independently surveyed. Electrophysiology, cardiomyopathy and cardiac transplant patients were excluded. Results were scored as full, partial or incomplete recognition.
Results: Most (98% parents; 83% adolescents) reported that their cardiologist had provided education. Summary results are noted in Table I. Older adolescents performed better. Parents performed better than all patients. History of prior surgery/intervention had no significance. Patients undergoing last surgery >11 y.o. scored better than those with surgery at younger age or without surgery ($p < 0.05$). Patients with complex lesions (single ventricle, tetralogy of Fallot, transposition of the great arteries) were better at naming their defect ($p < 0.05$) but similar in ability to describe the anatomy compared to those with minor lesions. Those with more complex disease were better at providing the full name of the defect (81/134; 60%) than in fully describing it (43/134; 32%; $p < 0.005$).
Conclusions: Unlike their parents, only 1/2 of adolescents can name their cardiac defect and 1/3 can adequately describe the anatomy. Older adolescents and those with more complex defects are more knowledgeable. A stronger emphasis should be made on educating this population.

Table 1.

	Name defect (n = 522)		Describe defect (n = 521)	
	Full	**Partial**	**Full**	**Partial**
11-14 y.o	40% (n = 102)*	13% (n = 34)	23% (n = 58)*	35% (n = 89)
15-17 y.o.	51% (n = 87)*	17% (n = 29)	35% (n = 56)*	37% (n = 63)
> 18 y.o	68% (n = 65)*	16% (n = 15)	54% (n = 51)*	31% (n = 29)
TOTAL	**49% (n = 254)***	**15% (n = 78)**	**32 %(n = 165)***	**35% (n = 181)**
Parent	78% (402/512)*	11% (n = 56)	61% (312/512)*	29% (n = 151)

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P1794 - PERCEIVED EFFECT OF CARDIAC DEFECT ON EXERCISE ABILITY AND QUALITY OF LIFE IN ADOLESCENTS

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Introduction: Exercise ability and quality of life (QOL) vary widely in patients with congenital heart disease (CHD), depending on the type of defect. It is expected that those with more severe CHD would be more adversely affected.
Methods: Adolescents with CHD were asked to describe how their CHD affected their exercise ability and how it would "affect your life as an adult?" Answers were scored as 'None,' 'Mild', 'Moderate' and 'Severe' restriction.
Results: The vast majority of patients expect no (63%) or minimal (28%) effect on long-term QOL. Single ventricle (SV) patients were most severely affected, followed by those with repaired tetralogy of Fallot (Table I). These findings correspond with perception of current exercise ability (Table II).
Conclusions: The vast majority of adolescents with CHD do not perceive that it significantly adversely affects their exercise ability or QOL. Patients with more severe lesions report more significant effect.

Table I. Perceived effect of CHD on QOL.

	Shunt (n = 131)	Left-sided (n = 168)	Tetralogy (n = 59)	TGA (n = 23)	SV (n = 37)	Total (n = 418)
None	80% (106)	60% (101)	41% (24)	70 % (16)	30% (11)	63% (258)
Mild	18% (23)	32% (53)	49% (29)	13% (3)	32% (12)	28% (120)
Moderate	2% (2)	6% (12)	10% (6)	17% (4)	32% (12)	8% (36)
Severe	0	1% (2)	0	0	5% (2)	1% (4)

Table II. Perceived effect of CHD on exercise ability.

	Shunt n = 136	Left heart n = 164	TOF n = 58	TGA, TAPVR n = 24	SV n = 40	Total n = 422
None	79% (n = 108)	66% (n = 109)	41% (n = 24)	54% (n = 13)	20% (n = 8)	62% (n = 262)
Mild	17% (n = 23)	25% (n = 41)	41% (n = 24)	17% (n = 4)	30% (n = 12)	25% (n = 104)
Moderate	3% (n = 4)	8% (n = 13)	16% (n = 9)	25% (n = 6)	45% (n = 18)	12% (n = 50)
Severe	1% (n = 1)	1% (n = 1)	2% (n = 1)	4% (n = 1)	5% (n = 2)	1% (n = 6)

P1797 - CLASSIFICATION OF CORONARY ARTERY ANEURYSMS IN KAWASAKI DISEASE BASED ON Z SCORE IN A MEXICAN POPULATION

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Background: Kawasaki Disease is the most common acquired heart disease in developed countries. Classification of coronary artery abnormalities (CAA) has been changed across the years. Recently the use of z-score based on body-surface-area has increased the accuracy of definition or coronary abnormalities.
Objective: Compare the definition of coronary artery abnormalities in KD using the classification based on coronary intraluminal diameter proposed by the American Heart Association with the z-score classification based on body-surface-area.
Material and Methods: All patients in the acute phase of KD diagnosed at the Instituto Nacional de Pediatría in Mexico City, Mexico from August 1995 to of August 31st, 2015 were included. Coronary artery luminal dimensions were converted to body-surface-area adjusted z-scores and compared with the classification proposed by Manlhiot. The Kappa correlation test was used to evaluate the accuracy of both classifications.
Results: A total of 384 patients with KD were reviewed. When we used the AHA classification based on coronary artery luminal dimensions, 141 cases were diagnosed with coronary artery abnormalities in the acute phase of KD (36%). When we adjust the classification of CAA using the body-surface-area 146 cases were diagnosed with CAA (37.6%). The main differences between these classifications were present in small children and in large coronary artery aneurysms. The AHA classification overestimated the severity of small CAA, but underestimated significantly in giant coronary aneurysms. (85.7%)
Conclusions: Classification of CCA based in body-surface z-score improves the correct classification of CAA in KD patients. This improvement is important for defining management especially in small children.

P1802 - FIRST DATA OF CLINICAL PRESENTATION OF SIBLINGS WITH DIAGNOSED MARFAN SYNDROME MANY BUT NOT ALL

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Objectives: Marfan syndrome(MFS) is a genetic disorder with a variable clinical presentation. Most serious manifestations may affect heart, aorta, skeleton and eyes. The aim of our study is the external validation of the clinical presentation of siblings with an identical evidence of MFS.
Methods: Since 2008 we investigated 381 patients (11.4 ± 5.5y) with suspected MFS, whereas 30 patients are siblings with a genetic (n = 21) or clinical evidence(n = 9) for MFS (13.0 ± 5.8 y; age difference of siblings: 3.7 ± 3.1 y; 18 male: 12 female). We analyzed most serious manifestations: dilation of sinus valsalvae (SV), atrio-ventricular-valve prolapse (AVP), ectopia lentis (EL) and skeleton indices (SK, ≥3), (n = 30). Manifestation of dural ectasia (DE) was included for patients who were suitable for MRI.
Results: In all manifestations except DE, siblings showed a high percentage of identical clinical presentation (SV 71%, AVP 86%, EL 86%, SK 71%, DE 50%). The impact of SV and SK was higher in the group with genetic evidence of MFS (56% vs 25% and 44% vs 25%) but was not influenced by any other parameter. EL was found in 50% of siblings with genetic evidence (clinical evidence 13%). In all siblings without genetic evidence an AVP in at least one sibling was found. The data of the 30 patients shows no influence of gender, age difference and period of observation.
Conclusion: We can conclude that siblings with diagnosed MFS showed a high percentage of identical clinical manifestation. There is an obvious different impact of clinical manifestation between siblings with or without a genetic evidence of MFS. The reason why some siblings with the same evidence for MFS showed a different clinical presentation is still unknown. Further investigations with a larger collective are needed to amplify the results. These findings may be relevant for the clinical guidance of affected families by allowing a better estimation of prognosis and individual therapy.

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P1818 - PAEDIATRIC PREVENTION OF ADULT CARDIOVASCULAR DISEASES IN THE UAE

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Background/Hypothesis: Atherosclerosis or cardiovascular disease (CVD) starts early in life. In children the disease is asymptomatic and characterized by a progressive initial phase that is hastened by serious health problems such as obesity, hypertension, diabetes and dyslipidemia. Early recognition and prompt intervention halt the progression of atherosclerosis in children with increased risk factors for developing CVD as adults.
Aims: To determine the prevalence and risk factors of obesity, hypertension, diabetes and dyslipidemia in Emirati children. These aims are purposed by identifying obese, hypertensive, pre diabetic diabetic and dyslipidemic students in the study cohort. Children with increased CVD risk will be followed for risk reduction and counselling.
Methodology: The study population n = 523 included school-age children at grades 2 elementary school, 6 middle schools and 10 high school of ?200 student's grade. Parents will complete the study questionnaire which addresses consanguinity and risk factors for cardiovascular disease. Participants had the following determinations: Height, weight body mass index, waist circumference, blood pressure, and pulse rate. Venous blood samples will be collected for measuring the lipid profile total cholesterol high density lipoprotein low-density lipoprotein, and triglycerides random blood glucose, hemoglobin A1c, adiponectin and inflammatory biomarkers interleukin6, high sensitivity C reactive protein and tumor necrosis factor-α.
Results: The results will identify prevalence and risk factors of cardiovascular disease in our young Emirati students. Recognized health problems will be amenable to treatment counseling and risk prevention.
Conclusions.

P1826 - ASSESSMENT OF SERUM VITAMIN D LEVELS IN CHILDREN WITH CONGENITAL HEART DISEASE

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Background: Congenital heart disease (CHD) is a relatively common problem with an estimated prevalence of 1 per 100 in the general population. A significant number of these patients will require corrective surgeries over their lifetime. Emerging literature suggests vitamin D deficiency to be a highly prevalent problem in the immediate post-operative CHD population. Also many studies have shown that vitamin D deficiency contributes to secondary organ pathophysiology and worsens outcome in critically ill populations. In this study we aimed to compare serum vitamin D levels in children with CHD who were candidate for corrective surgery with children without CHD.
Materials and Methods: In a case-control study that was conducted in 2015 in Mashhad /Iran, serum levels of vitamin D in 44 children with CHD was compared to 44 controls. Cases of congenital heart disease diagnosed by echocardiography and were recruited by convenience sampling. The control group was selected from healthy less than 2 year's old children who referred to health centers for routine care. Exclusion criteria for both groups were concurrent systemic diseases, irregular consumption of multivitamin drops and malnutrition. Data analysis was done in SPSS V 20 software and descriptive statistics, t-test and analysis of covariance were used.
Results: The mean age in cases was 10.5 ± 1.7months and in controls was 14.5 ± 3.6 months. In cases, 24 patients (54.5%) and in controls 22 cases (50%) were male. The mean serum vitamin D values in subjects 38.2 ± 30 ng/dl and controls 42.6 ± 18 ng/dl was significantly different ($P < 0.05$). Age and sex had no significant effect on vitamin D values.
Conclusions: There was a significant difference in serum vitamin D values in subjects and controls. Therefore, we suggest that the serum levels of vitamin D in children with CHD should be checked especially before corrective surgery of congenital heart disease.

P1836 - CARDIAC HEMANGIOMA IN INFANCY. A CASE OF SUCCESSFUL TREATMENT WITH PROPRANOLOL

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Hemangiomas are a type of vascular tumors that could be associated with morbidity and mortality in infants and children. Propranolol has been used successfully in treating patients with hemangiomas in different organs but its effectiveness has not been reported in cardiac hemangiomas. We present a case of an infant with a right atrial cardiac hemangioma treated with propranolol.
Case Report: 16 days old male newborn infant, born at term referred for cardiac evaluation of a holosystolic heart murmur with normal vital signs. Echocardiography showed a large, 33 by 16 mm, multiloculated mass, centrally heterogenous with well-defined edges extending from the roof of the right atrium to the tricuspid valve. Color Doppler demonstrated flow in the central lacunae. Obstruction of the SVC or IVC drainage into the right atrium was not seen and there was no interference with tricuspid valve function. Additionally, 2 apical VSD's 2 × 3 mm each were seen. Cardiac MRI confirmed the diagnosis. Treatment with oral propranolol was initiated at progressive doses up to 2.5 mg/kg/day. This was well tolerated without adverse effects. Follow up echocardiography showed a gradual decrease in tumor size and blood flow starting peripherally as early as 2 weeks after initiation of therapy. There was complete resolution of the tumor by echocardiography and MRI at the age of 1 year. The VSD's closed spontaneously at 18 months.
Conclusion: Propranolol was effective in treating this infant with a rare cardiac hemangioma. A quick favorable response was observed and, in the absence of significant hemodynamic consequences requiring surgical intervention, propranolol should be the first line of therapy in cardiac hemangiomas as well.

P1847 - PEDOMETER ACCURACY FOR MEASURING PHYSICAL ACTIVITY BEHAVIOUR DECREASES WITH AGE AMONG PAEDIATRIC CARDIOLOGY PATIENTS

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Background: Physical activity (PA) counselling is recommended at every clinical encounter for those with congenital heart defects.

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Clinicians recognize that self- or parent-proxy reports of patients' PA can be highly inaccurate. Pedometers are simple, inexpensive tools to objectively and reliably measure PA levels in adolescents and adults. This study evaluated whether pedometer accuracy varies by age, as PA patterns differ significantly between young children and adolescents.
Methods: A convenience sample of eligible participants (n = 22) between 3 and 14 years of age wore a belt with an attached pedometer and accelerometer for one week. Step counts and time spent in moderate to vigorous physical activity (MVPA) were compared between devices, accounting for age, sex and diagnosis.
Results: Accelerometer and pedometer step counts ($r = 0.76$) and MVPA ($r = 0.53$) were moderately correlated, with significant agreement (Kappa = 0.66), but pedometers overestimated activity relative to accelerometer results. The devices were most strongly correlated for steps ($r = 0.98$) and MVPA ($r = 0.97$) among adolescents (12 to 14 years old). The discrepancy in steps between devices was significantly greater for young children (3 to 5 years) compared to older children (6 to 11 years, $p = 0.02$) and adolescents (12 to 14 years, $p = 0.05$). Differences in time spent in MVPA were similar (greater discrepancy in younger children compared to older children [$p < 0.01$] and adolescents [$p < 0.01$]). Diagnosis and gender did not contribute to device differences ($p > 0.35$).
Conclusion: Pedometers are easy to use, inexpensive tools which can provide clinicians with reliable, objective information on the PA of adolescent patients. The large discrepancy between pedometer and accelerometer measures among younger children suggests that pedometers may not be as suitable for assessing their more diverse PA movement patterns.

P1862 - ORALLY ACQUIRED AND TREATED CHAGAS DISEASE AFTER 9 YEARS. IS IT ALL OVER

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Introduction: in Latin America a sparse number of orally acquired Chagas disease outbreaks have been reported. The largest urban outbreak occurred in a school in Caracas- Venezuela in 2007. In this event 103 people were infected by ingestion of guava juice prepared under inadequate sanitary conditions.
Objective: The aim of this study is to describe nine years of cardiovascular follow up in a group of patients with Chagas disease by oral transmission.
Methods: Since 2007 we followed 22 pediatric patients from an outbreak of oral acquired Chagas disease. The annual evaluation consists in a clinical exam, 12-lead electrocardiogram, Doppler echocardiogram, 24 hour Holter monitoring and serologic tests.
Results: Of the 103 patients, 22 had been in cardiovascular control (59% males, 41% females) with a mean age of 16,22 ± 3,81, weight of 57,07Kg ± 16,77 and size of 164,45 cm ± 12,59. 100% of the patients received treatment with Nitro derivates compounds, 50% required a second cycle between 2010-2011 and 4,5% required a third cycle in 2012. Among the electrocardiographic findings, 50% of the patients had early repolarization disorders, 18% intraventricular conduction disorders, 18% sinus bradycardia, and 4,5% incomplete right bundle branch block. Echocardiographic findings indicate that 45% of patients had a restrictive diastolic filling pattern of LV, 36% of RV and 9% already show an increase in the final diastolic diameter of LV. In the Holter monitoring, < 3,5 sec sinus pauses were found (18%), low atrial rhythm (13,5%), 1st degree AVB (4,5%), premature supraventricular beats (13,5%) and premature ventricular beats (4,5%)
Conclusion: Persistent serological changes in patients with acute oral Chagas with adequate treatment suggest a progression of the disease. Despite of it, we strongly suggest lifetime follow up ptotocol including electrocardiographic and echocardiographic evaluations as a prognostic tool of chronic cardiomyopathy development.

P1916 - ISOLATED AORTIC DILATION WITHOUT OSTEOARTHRITIS A CASE OF SMAD3 MUTATION

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Introduction: Thoracic aortic aneurysms and dissections (TAADs) are often found in the context of genetic syndromes. Aneurysms-osteoarthritis syndrome (AOS) is a recently discovered inherited autosomal dominant connective tissue caused by SMAD3 mutations. AOS responsible for 2% of familial TAADs and characterized by aneurysms, dissections and tortuosity throughout the arterial tree in combination with osteoarthritis. Early-onset osteoarthritis is present in almost all patients. We present a patient with aortic root dilation caused by pathogenic mutation in SMAD3 gene with atypical clinical presentation.
Case Report: Asymptomatic 11 years old male, referred to our department with dilation of the aortic root diagnosed. Neither craniofacial abnormalities such as hypertelorisim and bifid uvula, nor chest deformities were found. Echocardiography revealed dilation of the aortic root and ascending aorta that rapidly progressed in subsequent follow-up echocardiograms. Imaging assessment was completed with MRI. Surgical repaired was deemed necessary and patient underwent a Bentall procedure with good outcome. Genetic study showed pathogenic mutation on SMAD3 (Alteration c.733 G > C (p. Gly245Arg)). Neither joint abnormalities nor cerebrovascular tortuosities were found on complimentary examinations.
Discussion: We describe a patient with AOS with typical cardiovascular findings but without joint abnormalities. Identification of the underlying genetic defect in TAADs is crucial, considering the variability in prognosis, treatment strategy, and risk assessment in family members. Cardiologists should suspect AOS in all patients with aneurysmal dilation of the aorta of unknown cause. Early

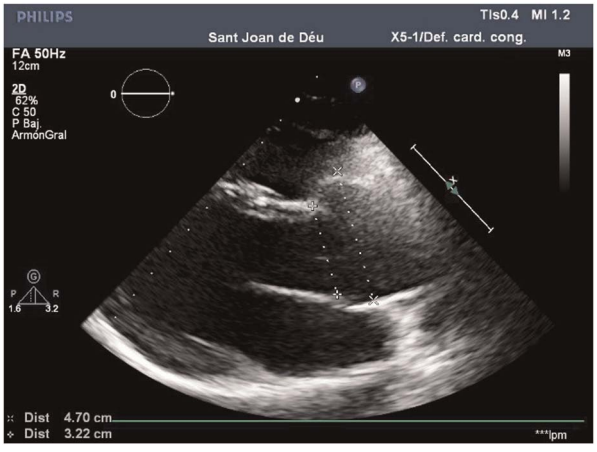

Figure 1.

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elective surgical intervention is indicated in AOS patient's because dissections can occur in relatively small aortic diameters. A multidisciplinary approach including clinical geneticists, cardiologists, orthopedic surgeons, radiologists, neurologists, and vascular or cardiothoracic surgeons when necessary is key to manage the multisystem involvement of these patients

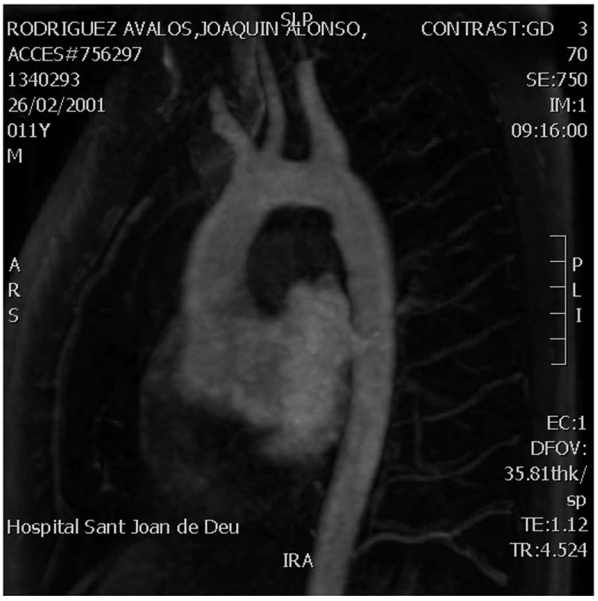

Figure 2.

P1942 - THE EFFECT OF PHYSICAL ACTIVITY BOUT PATTERNS ON BLOOD LIPIDS IN YOUTH NHANES 2003 2006

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Background/Hypothesis: Some evidence suggests that moderate to vigorous physical activity (MVPA) accumulated in bouts of 10 continuous minutes or longer have an independent effect on body composition, cardiorespiratory fitness, bone density, and weight loss. Although the effects of physical activity (PA) bout patterns in adults is well documented, the effects of PA bout patterns on cardiometabolic risk in youth needs further study. The purpose of this study is to determine the effects of PA bout patterns on blood lipids and body composition in youth.
Materials/Methods: Data from children and adolescents (2-17 years of age; N = 1066) with a minimum of four valid days of accelerometer wear time participating in the National Health and Nutrition Examination Survey 2003-2006 were used. Accelerometers were assessed for time spent in sporadic bouts (<5 minutes) of MVPA, short bouts (5-10 minutes) of MVPA, and medium-long bouts (≥10 minutes) of MVPA. Cluster analysis grouped subjects into three distinct exercise patterns. Covariate adjusted general linear models were used to compare blood lipid and body composition outcomes of individuals of different exercise patterns.
Results: PA pattern characteristics are as follows: Cluster 1 (96.6% sporadic, 2.5% short, 1.0% medium-long bouts, n = 718); Cluster 2 (89.1% sporadic, 8.8% short, 2.0% medium-long bouts, n = 232); and Cluster 3 (82.7% sporadic, 10.9% short, 6.3% medium-long bouts, n = 115). After controlling for age, sex, BMI percentile, and total MVPA minutes, there were no differences between clusters for total ($p = 0.877$), HDL ($p = 0.410$), or LDL ($p = 0.753$) cholesterol, triglycerides ($p = 0.295$), systolic ($p = 0.287$) or diastolic ($p = 0.108$) blood pressure, waist circumference percentile ($p = 0.145$), waist to height ratio ($p = 0.793$), or body composition ($p = 0.631$).
Conclusions: MVPA bout patterns, independent of total MVPA minutes, have no effect on blood lipids or body composition in children. Youth should be encouraged to accumulate the recommended minutes per day of MVPA without regulation for pattern or bout duration.

P1945 - HYPERTRIGLYCERIDEMIC WAIST PHENOTYPE PREDICTS VASCULAR HEALTH IN YOUTH WITH DYSLIPIDEMIA

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Background/Hypothesis: The hypertriglyceridemic waist phenotype (HTWP) is defined by the presence of elevated waist circumference (WC) (≥90th percentile) combined with elevated triglycerides (≥1.1mmoL). Linking WC with triglyceride concentrations allows clinicians to stratify presence of metabolically active visceral fat opposed to less metabolically active subcutaneous fat. In youth, the HTWP is associated with low fitness, elevated total cholesterol, low HDL cholesterol, impaired fasting glucose, and high diastolic blood pressure. However, the effects of the HTWP and this clustering of risks on vascular health in children is unknown. The purpose of this study is to determine the effect of the HTWP on vascular health in youth with dyslipidemia.
Materials/Methods: Children and adolescents aged 8-15 years were recruited from a pediatric preventive cardiology clinic. All participants completed a full blood lipid panel. WC was measured 2 cm above the iliac crest by a single observer. Pulse wave velocity (PWV) and augmentation index (Aix) were measured using a combined oscillometric and tonometric device. Carotid artery intima-media thickness (CIMT) was measured on the far wall of the left and right common carotid arteries using a semi-automated edge detection software.
Results: Of the 115 youth (males = 58; mean age = 13.05 ± 1.86 years), 65 met criteria for HTWP. After controlling for blood pressure, LDL and HDL cholesterol, children with HTWP had an elevated Aix (standard) (HTWP 14.43% ±1.74; normal 7.96% ±2.04; $p = 0.025$), Aix (heart rate controlled at 75bpm) (HTWP 11.91% ±1.77; normal 5.41% ±2.08; $p = 0.027$), and CIMT (HTWP 0.466mm ±0.004; normal 0.468mm ±0.005; $p = 0.821$). There were no differences in PWV (meter/sec) (HTWP 5.19 ±0.114; normal 5.01 ±0.128; $p = 0.327$).
Conclusions: Presence of the HTWP is associated with vascular damage characterized by increased Aix in children with dyslipidemia. HTWP can be used to stratify children who are at increased risk for premature vascular disease.

P1948 - IMPACT OF INTRAUTERINE GROWTH RESTRICTION ON CARDIOPULMONARY EXERCISE DURATION

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Background: Infants subjected to impaired delivery of nutrients and oxygen are at risk for impaired fetal growth, commonly termed

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intrauterine growth restriction (IUGR). Multiple studies have demonstrated association of IUGR with early and late-onset cardiovascular pathologies, including systolic and diastolic dysfunction, altered sarcomere structure, decreased cardiomyocyte endowment, increased cardiac fibrosis, hypertension, and adult-onset ischemic heart disease. The effects of these changes on exercise capacity in children and adolescents are unknown.
Methods: We performed a retrospective chart review of a single-institution's cardiopulmonary exercise stress lab database on patients born after 1/1/1990 with cardiopulmonary exercise testing (CPET) prior to 7/1/2016. We cross-referenced documented birth weight and gestational age to obtain a weight-for-gestational age percentile. Individuals with birth weight <10th percentile for gestational age were defined as IUGR. We excluded patients with a known history of congenital heart disease or cardiomyopathy. Normal controls were obtained from a previously defined cohort of healthy patients at Children's Hospital of Wisconsin. Controls were matched 2:1 with IUGR cases by gender and age at CPET.
Results: A total of 126 patients were included with 42 IUGR cases and 84 controls. IUGR patients had mildly decreased exercise duration that did not reach significance (11:54 versus controls 12:34, $p = 0.14$). There was no significant difference in heart rate response (IUGR 114.5 ± 17.2 bpm; controls 119.7 ± 17.0, $p = 0.12$). Regression analysis confirmed that IUGR was not an independent predictor of exercise duration but age, gender, and BMI were predictors.
Conclusions: IUGR is not associated with significant differences in measurements of exercise capacity in children, though there was a mild difference in exercise time. This study was limited by patient numbers. Further research is needed to study the effects of the pathophysiology of growth restriction on cardiovascular and exercise physiology.

P1964 - NEURODEVELOPMENTAL OUTCOMES IN CHILDREN WITH SINGLE VENTRICLE CIRCULATION UNDER THE AGE OF FIVE YEARS RECRUITED IN THE LONDON AREA

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Background: Neurodevelopmental abnormalities are common in children with heart disease, with the greatest risk in children with single ventricle (SV) circulation. There is no current process for routinely monitoring this patient group in the UK, resulting in late detection and referral.
Methods: Children with SV circulation under the age of 5 years were recruited from three children's heart centres in London between January 2014 and July 2015 as part of a wider cross sectional study. Neurodevelopment was evaluated with the Mullen Scales of Early Learning (MSEL) cognitive score (all children) and MSEL motor score in children aged <33 months. Parents completed the Ages and Stages questionnaire (ASQ-3).
Results: Ninety patients with SV circulation with a median age of 15 months were evaluated, 52 (58%) were male, 44 (49%) had HLHS, 46 (51%) other single ventricle lesions. Fifty-five patients (61%) had undergone more than 1 congenital heart operation and 18 (20%) had a non cardiac diagnosis linked to developmental delay. Of 90 children assessed with MSEL for cognitive function, 18 (20%) had scores 1-2 SD below and 10 (11%) had scores >2 SD below the normative mean. Of 61 children assessed with MSEL for gross motor function, 12 (20%) had scores 1-2 SD below and 8 (13%) had scores >2 SD below the normative mean. Parents of 80 children completed the ASQ, with 19 (24%) obtaining borderline and 45 (56%) abnormal results.
Conclusions: A higher proportion of children with SV circulation assessed by the MSEL have impaired neurodevelopmental outcomes compared to the normal population. The ASQ flagged up a much higher proportion as abnormal than the MSEL, and may be picking up a wider range of health related issues. There is a need for improvement in the quality of neurodevelopmental assessment for children with SV circulation in the UK.

P1993 - CARDIOPULMONARY EXERCISE TESTING IN BOYS WITH ATTENTION DEFICIT HYPERACTIVITY DISORDER

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Background: Exercise capacity is one of the most important indicators of health, in children with attention hyperactivity deficit disorder (ADHD) physical activity increases their concentration, cognition, improves motor behavior and exercise tolerance. The aim of the study was to evaluate the exercise capacity of boys with ADHD.
Methods: Cardiopulmonary exercise test (CPET) was conducted in 37 boys with ADHD, (7-17 years, mean 11.3 ± 2.6) who were divided into 3 age subgroups: I – 7-10 years, mean 9.0 ± 0.3 years, II – 11-13 years, mean 12.1 ± 0.2 years , III – 14-17 years, mean 15.5 ± 0.4 years. Their results were compared with two age-matched reference groups REF1 ($n = 68$, 8-19 years, mean 14.2 ± 2.9) and REF2 ($n = 69$, 6-17 years, mean 11.5 ± 3.5) of healthy boys.
Results: ADHD boys achieved significantly lower workloads (116 ± 7 W) than the healthy boys (REF1-150 ± 8 W, $p < 0.01$) in the whole group, as well as in the age subgroups (I – 81 ± 6 W vs. REF1 118 ± 9 W; II – 114 ± 5 W vs. REF1 140 ± 7 W, both $p < 0.01$ and III – 175 ± 10 W vs. REF1 220 ± 12 W, $p < 0.05$). Peak VO2 in ADHD (1.56 ± 0.09 l/min) boys was similar to the REF1 values (1.62 ± 0.08 l/min), but significantly lower than in REF2 (1.92 ± 0.11 l/min, $p < 0.05$). Differential analysis in subgroups showed statistically significant difference only in subgroup II (1.41 ± 0.08 l/min vs 1.81 ± 0.09 l/min in REF2, $p < 0.01$). Anaerobic threshold was achieved at significantly lower workloads (subgroup I 50 ± 3 W vs. REF1 86 ± 5 W, $p < 0.001$; subgroup II 72 ± 7 W vs. REF1 91 ± 8 W, $p < 0.001$). No significant difference in subgroup III was found.
Conclusions: Decreased exercise tolerance was observed in boys with ADHD before their puberty. They reached the anaerobic threshold earlier than their healthy peers, thus the effort is performed less efficiently and associated with greater physiological strain. Cardiopulmonary exercise testing is a safe and valuable diagnostic tool in boys with ADHD.

P2047 - CHROMOSOME 22Q11.2 DELETION SYNDROME ANALYSIS OF ASSOCIATIONS BETWEEN CARDIAC AND EXTRACARDIAC MALFORMATIONS IN A COHORT OF 276 PATIENTS

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Cardiac malformations in 22q11.2 microdeletion syndrome (22q11DS) are frequent. The most common congenital heart disease (CHD) involve conotruncus and aortic arch. Extracardiac anomalies are also common. CHD, in association with other malformations may help suspect the diagnosis.
Objectives: Describe CHD in a cohort of Chilean patients with 22q11DS. Analyze their frequency and compare them with non-syndromic CHD population reported in literature. Assess the association between cardiac and non-cardiac malformations to search those suggestive of the syndrome.
Materials and Methods: review of clinical and demographic database of patients with 22q11DS diagnosed between 1998 and 2016.
Results: 276 patients; 148 (52.9%) were female. CHD was present in 50.4%. The most frequent were: VSD in 23.6%, ASD 14.3% , Tetralogy of Fallot (TF) in 18.9%; truncus arteriosus (TA) 3.2%; double outlet RV 1.8% and interruption of the aortic arch type B (IAA-B) 5.4%. With respect to extracardiac malformations, palate anomalies were present in 63.6%; CNS in 11.1%, renal 6.1%; skeletal 24.6%. According to Hoffman et al (2002), the frequency of CHD in the general population is 0.9%, with a frequency of VSD of 0.35%; ASD 0.09%; TF 0.04%; TA, 0.01%; DSVD 0.01% and IAA-b of 0.001%. No association of intracardiac malformations with aortic arch anomalies or between CHD and other malformations was observed in our cohort.
Conclusions: In this large, unselected cohort of patients with 22q11.2DS, the frequency of CHD was 50 times greater than that described for general population. It was 5,400 times more common for IAA-b B, 470 for TF and 60 for VSD. These results show the high risk of CHD in 22q11DS, and suggest that the deletion should be suspected in interruption of arch aortic and conotruncal heart disease per se, even in the absence of other anomalies. Fondecyt grant 1130392

P2117 - LATE EVALUATION OF PATIENTS UNDERGOING TOTAL CAVOPULMONARY BYPASS CLINICAL ASPECTS REINTERVENTIONS AND COMPLICATIONS THAT INTERFERE WITH MORBIDITY AND MORTALITY

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Introduction: The single ventricular hearts (SVH) correspond to a group of congenital heart diseases with several anatomical combinations. The treatment is performed through surgical staging culminating in the realization of Total Cavopulmonary Shunt Operation (TCSO) as final stage.
Objective: To evaluate the delayed evolution of patients submitted to TCSO in relation to complications thrombosis, protein-losing enteropathy, ventricular dysfunction and hepatic cirrhosis) and the need for reintervention, relating possible factors that may interfere in morbidity and mortality. Material and method: a cross-sectional study of patients submitted to TCSO with an extracardiac conduit (EC), from Jun/2003 to Jan/2016, in outpatient follow-up. Data obtained by survey of medical records. Descriptive analysis was performed and retrospective analysis of age at the time of surgery, presence of fenestration in the EC, perioperative thrombosis and previous pulmonary pressure.
Results: A total of 59 patients were analyzed, (70.2% males). Mean age at surgery: 4.45 ± 2.69 years. Left single ventricular heart performed 54.4%. Average follow-up time approximately 49 months. 68.4% had a mean pulmonary artery pressure (PAP) lower than 15 mmHg. 61.4% did not undergo EC fenestration. 35.1% had thromboembolic complication. When comparing the presence of thrombus 50% use of ASA. There was no statistical significance in the relation of the presence of fenestration, Left Heart Hypoplasia Syndrome and PAP > 15 mmHg with the appearance of protein-losing enteropathy. 15% had ventricular dysfunction, and 77% of then were Right Single Ventricle (RSV). 50% had some degree of hepatic congestion/hepatomegaly. 21% required re-interventions. No patient presented with plastic bronchitis or arrhythmias.
Conclusion: There was no statistical significance between patients who used ASA or oral anticoagulant for antithrombotic prophylaxis. The only preoperative factor that interfered with late evolution was RSV.

P2182 - PALIVIZUMAB IN INFANTS LESS THAN 1 YEAR WITH HEMODYNAMICALLY SIGNIFICANT CONGENITAL HEART DISEASE IN ARGENTINA A COMPARATIVE STUDY WITH HISTORICAL CONTROL GROUP

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Background: In 2009, Argentina launched a national program to assist children with congenital heart disease (CHD) and, since 2014, Health Ministry provides palivizumab (PVZ) to all infants with CHD. The Hospital Nacional de Pediatría Garrahan is a tertiary center with a Follow-up Clinic for children with chronic medical conditions (CMC). The study main objective was to analyze the impact of immunoprophylaxis with PVZ, comparing a cohort of infants with hemodynamically significant CHD (HS-CHD) that received PVZ vs. a historical cohort of HS-CHD infants not receiving PVZ.
Subjects and Methods: Prospective cohort study with historical control. HS-CHD patients <1 year of age, followed up at the CMC clinics between 2014 and 2016 were included. All patients received PVZ according to local guidelines. HS-CHD was defined with usual criteria. Historical control group comprised HS-CHD patients <1 year of age, followed up between 2007 and 2009, not receiveing PVZ. Data corresponding to respiratory syncytial virus (RSV) high circulation months (May to September) were analyzed. Demographic and clinical data, global hospitalization rates, RSV-specific hospitalization rates, length of hospitalization and need for mechanical ventilation were compared. Descriptive statistics, t-test, Wilcoxon test and chi-square test were used as appropriate. Differences are expressed as Relative Risk (RR), RR reduction, CI95% and Number Needed to Treat (NNT). A $p < 0.05$ was considered statistically significant.
Results: Fifty-three patients were included in PVZ group and 50 patients in control group. Table 1 shows groups characteristics. There were 10 respiratory tract infection hospitalizations (19%) in PVZ group vs. 15 (30%) in control group. Forty per cent of all hospitalizations were in June. There were no respiratory deaths. Table 2 shows differences between groups.

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Conclusions: RSV-related hospitalization rate was significatively lower in PVZ group. PVZ impact was high, even though patients with severe HS-CHD were more frequent in PVZ group.

Table 1. Groups characteristics.

	PVZ Group (n = 53)	Control Group (n = 50)	P
Follow-up period	2014-2016	2007-2009	
Sex (M/F)	27/26	33/17	0.08
Birth weight (g) (x ± DS)	3,240 ± 460	3,077 ±590	0.29
Gestational age (w) (x ± DS)	39.0 ± 1.3	39.0 ± 1.7	0.89
HS-CHD classification:			
Gl*	14 (26%)	17(34%)	0.27
G2*	31 (59%)	16(32%)	0.006
G3***	8 (15%)	17 (34%)	0.02
Single Ventricle/LVH	27 (51%)	11(22%)	0.001
Pulmonary hypertension	18 (34%)	9 (18%)	0.02

PVZ: Palivizumab. **HS-CHD**: Hemodynamically significant congenital heart disease. ***Gl**: Left-to-right shunt with congestive heart failure. ****G2**: Cyanotic CHD uncorrected or palliated (including single ventricle lesions). *****G3**: Corrected **CHD** with residual defects. **LVH**: Left ventricular hypoplasia.

Table 2. Outcomes.

	PVZ Group (n = 53)	Control Group (n = 50)	p
Follow-up period	2014-2016	2007-2009	
Age at hospitalization (months) median (range)	6.5 (3.0-9.0)	6.0 (4.0-6.0)	0.39
Length of stay (days) median (range)	6.0 (5.0-12.0)	7 (4.0-15.0)	0.23
RTI hospitalization (global)	10 (19%)	15 (30%)	0,19 RR: 0,63 (0,21-1,27) NNT: 8,98
RSV-related RTI hospitalization	3 (6%)	10 (20%)	0,04 RR: 0,28 (0,08-0,97) NNT: 6,97
Mechanical ventilation	2 (4%)	4 (8%)	0.32 RR: 0,47 (0,09-2,46) NNT: 23,6

PVZ: palivizumab. RTI: Respiratory tract infection. RSV: Respiratory Syncytial Virus

P2228 - IMPACT OF REAL TIME TELECONSULTATIONS IN PEDIATRIC CARDIOLOGY ELEVEN YEARS OF EXPERIENCE

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Background: A telemedicine program was initiated between the Pediatric Cardiology Department of a tertiary care hospital in Barcelona and pediatric cardiologists from 2 remote hospitals (around 100 km distance) in 2006 and 2008 respectively. The Project consists of real-time teleconsultation with direct connection of the echocardiogram images in real-time and in presence of the family (seeing people face to face), using a videoconference system with internet protocol (IP) connectivity with encrypted information.
Methods: We performed a retrospective review of all teleconsultations from 2006 to 2016 and analyzed the population, indications for teleconsultation, diagnoses, impact and satisfaction of the families (obtained through a satisfaction survey carried out in the first 118 teleconsultants).
Results: Over ten years, a total of 515 real-time videoconference were carried out, 388 with the first Hospital and 127 with the second, corresponding to structural congenital heart disease (97,2%) or arrhythmia disorders (2,8%); 220 (42,7%) females and 295 (57,3%) males; 9,9% were neonates. The most frequent reason of connections was cases for surgery 157 (30,5%), followed by the evaluation of children and follow-up 148 (28,7%), patients to plan an interventional catheterization or electrophysiology study 146 (28,4%) or second opinion (12,4%) (see Figure). The satisfaction of parents (118 patients) was very high in the survey (9.5 over 10) and a tele-consultation was preferred more than traditional consultation in 99,2% of cases. There were minor discrepancies between pediatrics cardiologists in 9,7% of the cases.
Conclusions: In our experience, real-time telemedicine is an important tool in the diagnosis and follow-up of children with cardiovascular diseases. It plays an important role in continuous medical training for the staff of hospitals in remote areas of a tertiary hospital and has significant medical, economic and social benefits for patients and their families.

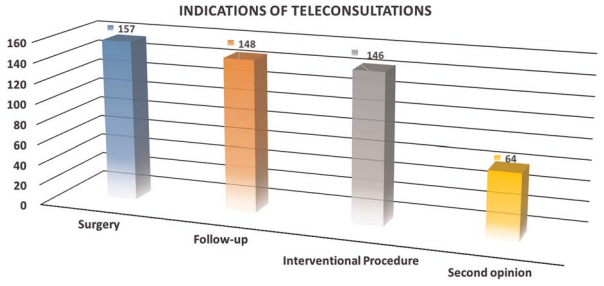

Figure.

P2238 - KAWASAKI DISEASE MARRAKECH UNIVERSITY HOSPITAL EXPERIENCE

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Introduction: Kawasaki disease (KD) is the leading cause of acquired heart disease among children which affects the coronary arteries. We aimed to evaluate the clinical manifestations, laboratory parameters, echocardiographic findings at diagnostic and assess the follow-up of KD in children.
Patients and Methods: In this retrospective study, we studied the clinical data, of all children with KD who had been admitted in the infectious pediatric department at Mohamed VI University Hospital Marrakech, from January 2011 to November 2016.
Results: The study enrolled 50 patients. Sex ratio was 1.5. The age at the disease onset ranged from 4 months to 12 years. All patients had a persistent fever, followed by oral mucosal lesions. 45 patients had typical KD, 5 had atypical KD. All patients had elevated CRP, 95% had elevated ESR. Elevated leukocyte count was observed in 60% and thrombocytosis in 40% of the patients. 42% of the

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echocardiography was performed during the acute stage of the disease. There were 16 cases , with lesion of coronary artery; pericarditis in two patients and an aneurysm of the left humeral artery in one child. During the follow-up, cardiac involvement was documented in 32 cases; coronary artery dilatation resolved in 2 children, the patients that were not found with arterial coronary at the acute stage had no abnormality in the follow-up. 44 patients received intravenous immunoglobulin (IVIG). One patient was complicated by pleuresia and only one patient relapsed and died with sepsis and intestinal necrosis.
Conclusion: Different facts play a role in the diagnostic and management of KD in our context. There is a shortage of pediatric cardiologists in our university hospital, which drains all the cardiac children of the southern regions of Morocco. The expensive cost of IVIG is an obstacle for patients to access therapy.

P2240 - HOSPITAL PREVALENCE OF CONGENITAL HEART DISEASE IN A MOROCCAN CITY MARRAKESH

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Background: Congenital heart disease englobes all cardiac and vascular malformations. It is a major health problem, especially in developing countries. The aim of this work is to assess the prevalence of congenital heart disease.
Methods: It is a retrospective, descriptive and analytical study, including all children aged 1 to 18 years hospitalized in the pediatric department of Mohammed VI university hospital in Marrakech, with diagnosis of congenital heart disease, between January 2011 and November 2016.
Results: Out of 7627 patients admitted, 247 had a congenital heart disease, corresponding to a hospital prevalence of 3.2%. Mean age was 15.2 months. Boys represented 55.8% of cases. Children with Down Syndrome represented f25.9%, parental consanguinity represented 15.38%. concerning the diagnosis, VSD was the most frequent with 108 cases (43.7%), followed by ASD in 97 patients (39.2%), the PDA in 34 cases (13.7%), tetralogy of Fallot in 20 patients (8%), pulmonary stenosis in 16 cases (6, 4%), transposition of great arteries in 12 cases (4.8%). In children with trisomy 21, the most common malformation was AV canal, found in 25 patients (39%).
Conclusion : The prevalence of congenital heart disease is variable, could be underestimated in this study. The prevalence is constantly increasing, which could be explained by the development of investigation techniques but also lifestyle modification with more exposure to teratogenic risks. The high rate of consanguinity in Morocco exposes to even more congenital heart disease. The lack of resources is the major problem of management in our context. Knowledge of the prevalence of congenital heart disease could allow for good organization and planning of care, from diagnosis to treatment.

P2256 - CLINICAL CHARACTERISTICS COMPARISON BETWEEN CLASSICAL AND ATYPICAL KAWASAKI DISEASE IN UNIVERSITY MALAYA MEDICAL CENTRE

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Introduction: Kawasaki disease (KD) is a self limiting vasculitis commonly affecting children less than 5 years old. We aimed to study the clinical characteristics between classical and atypical KD by examining the coronary artery (CA) status and systemic inflammatory markers.
Methods: A single centre, retrospective review of children with KD during 4 years. All children had an echocardiography at diagnosis, and at 1,3 and 6 months after diagnosis. Patients received intravenous immunoglobulin (IVIG) and antiplatelet therapy after confirmed to have KD.
Results: A total of 43 children with mean age 23.9 ± 18.2 months, 23 male and 20 female were included. KD were of classical type in 33, 8 atypical and 2 incomplete type. Clinical characteristics includes fever duration of 6.65 ±2.09 days, cervical lymphadenopathy in 37(86%), polymorphous rash in 36(83.7%), non purulent conjunctivitis in 34(79.1%), erythematous lips in 37(86%), extremities changes in 22(51.2%)and BCG scar flare in 16(37.2%). IVIG administered at 7.18 ± 2.18 days of illness. Atypical KD are more likely in children less than 6 months. Parameters includes hemoglobin, erythrocyte sedimentation rate and serum albumin is significantly elevated in classical compared to atypical KD with p value of 0.011,0.034,0.004 respectively. Coronary artery aneursyms commonly involve left main coronary artery(LMCA). At diagnosis, 10 patients had z score >2.50 for LMCA, 4 patients had z score >2.50 for right coronary artery (RCA) and 4 patients had involvement of both LMCA and RCA.6 patients had an increase in z score of LMCA at 1 month after diagnosis compared to at diagnosis.
Conclusion: Inflammatory markers are a useful surveillance markers in classical KD. Coronary artery aneurysm are less severe in classical KD.

P2364 - INCIDENCE OF CONGENITAL HEART DEFECTS IN CHILDREN WITH CHROMOSOME ABNORMALITIES IN SOUTH EASTERN EUROPEAN COUNTRY A POST WAR PERIOD

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Background: Congenital heart defects (CHD) etiologically are result of genetic aberrations and environmental factors. Chromosome abnormalities are one of genetic factors that come with a significant risk of CHD. Additionally, it is already known that maternal stressful life can increase prevalence of CHD in offspring.
Aim: Analysis of incidence and pattern of CHD in children with chromosome abnormalities syndromes.
Patients and Methods: Study has been conducted in post-war period 2007-2015 yr, in the University Clinical Hospital Mostar, Bosnia and Herzegovina. Altogether 62 children were diagnosed with numerical or structural chromosome abnormalities - syndromes, majority with Down (50), and rest with Edwards (4), Turner (2), Patau (2), DiGeorge (2), Wolf-Hirschhorn (1), Kleefstra (1). From the medical documentation two main findings were analyzed - cytogenetic and echocardiographic, and four additional parameters - gender, birth weight, birth term and outcome. Children were followed-up to 1 year old.

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Results: Of all patients, 51.6% (32/62) had CHD. The most frequent defects were as follows: AVSD (37.5%), VSD (21.9%), clinically significant ASD/PDA (15.6%), valvular defects (9.4%). Incidence of CHD in patients with certain syndromes were: in Down 46% (23/50), in Edwards 100% (4/4), in Turner 50% (1/2), and in other rare syndromes pulled together 66.7% (4/6). In Down syndrome the most common CHD was AVSD (12/23), in Edwards VSD (1/4), in Turner valvular defect, and in group of patients with other rare syndromes VSD and aortic coarctation/stenosis (1 and 1 out from 4, respectively). CHD incidence was higher in hypothrophic patients (87.5%) and generally led frequently to fatal outcome (7/32).
Conclusion: Knowing the incidence of CHD in specific syndromes has practical implications and is important in diagnostic chain. Our results of CHD incidence in patients with Down and Edwards syndromes are in accordance with the literature data, as well as the mortality rate.

P2375 - LRP2 AN ENDOCYTIC VESICLE TRAFFICKING PROTEIN ASSOCIATED WITH CONGENITAL HEART DISEASES

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Objectives: Congenital heart diseases affect nearly 1% of neonates per year in the United States and are the leading cause of neonatal death. Despite a strong indication of genetic contribution to congenital heart diseases, the developmental processes and genetic components are poorly understood. From the large-scale forward genetic screen using inbred mice chemically mutagenized with ethyl-nitrosourea (ENU), a finding of endocytic trafficking gene mutations, including Lrp2, were recovered as causing congenital heart disease phenotypes. Endocytosis plays an important role in modulating cell signaling by regulating the internalization, recycling and degradation of receptor-ligand complexes. This involves the regulation in trafficking of the internalized endocytic vesicles to different endosomal compartments, some destined for fusion with lysosomes and degradation while others are recycled to the surface by recycling endosomes. Mutations in LRP2 causes a spectrum of congenital heart disease comprising of outflow tract abnormalities and alignment defects including truncus arteriosus with/without aortic arch anomalies.
Methods and Results: Here we report the LRP2 [line 1625(MGI 5489925)] mutant mouse models recovered from a forward genetic screen. Exome sequencing identified line1625 (MGI5489925) results in an A to G substitution at nucleotide -3 of an intron before nucleotide 8456 (c.8456-3 A > G, NM_001081088), a presumed splicing mutation. The mutants exhibit a spectrum of septation and outflow tract defects including truncus arteriosus type II with or without aortic arch anomalies. we use a series of tissue-specific genetic manipulations to define the crucial role of the LRP2 pathway in these three fields of cells (endocardial cushion, secondary heart field and cardiac neural crest) during outflow tract development and septation. LRP2-Mef2C and LRP2-Nkx2.5 recapitulate LRP2m/m cardiac phenotype (truncus arterious with or without arch anomalies).
Conclusions: We provide the evidence of endocytic vesicle trafficking pathway associated with the development of congenital heart diseases with malalignment/sepetation and outflow tract defects

P2393 - HOME AED PREVENT SUDDEN DEATH IN CHILDREN !

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Introduction: Sudden death of children may occur at any time. Once the accident happens, it influences the life of family members, and also affects all concerned in the school and in the community. Automated external defibrillator (AED) was initially installed for general citizens in 2004 in Japan, resulting in an increased number of live people rescued by non-medical workers. Recently, it has been proven that sudden death in children is often caused by ventricular fibrillation, which is an indication of AED. However, providing the Home AED is still not common. Here, we report a case that has been life-saving by Home AED and the effectiveness and necessity of installation are discussed.
Case Report: A 13-year-old boy who has been emergency admitted to our hospital due to ventricular tachycardia.10 years ago, he was suffering from fulminant myocarditis complicated with refractory ventricular tachycardia. As a result, remained ventricular aneurysm as sequelae. Although recent clinical course is stable and the arrhythmia was not observed, this time when he was down the stairs of his house, palpitations happened suddenly, and was deteriorating rapidly. His mother noticed tachycardia, attached AED to him in great haste. AED instructed to shock of electrical cardioversion, she press the shock button without hesitation.
Discussion and Conclusion: The analysis results of the recording of AED demonstrated heart rate of 260 beets/min of ventricular tachycardia, if appropriate management is not performed by the mother, there is a high possibility that had led to death. Home AED is one of the effective methods in order to protect the children from sudden death. Especially having an underlying disease, it is necessary to consider aggressively introduced.

P2409 - CLINICAL SIGNIFICANCE OF CARDIAC BIOMARKERS IN CHILDREN WITH HEART PROBLEMS

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Background: Cardiac biomarkers are indicators of heart disease severity. B-type natriuretic peptide (BNP) or N-terminal pro B-type natriuretic peptide (NT-proBNP) are produced in the cardiac ventricles and are primarily used to evaluate the severity of heart failure. Cardiac troponin T (cTnT) is a specific marker of acute heart damage. Creatine kinase (CK) is relatively low specific for myocardial injury.
Aim: Aim of the study was to determine the level of NT-proBNP, cTnT and CK in children with various cardiac diseases.
Patients and Methods: Prospective analysis included 65 children with heart problem (30 male and 35 female) from 1 month to 17 years of age (mean 4 years and 7 months). They were divided into three groups: 1. native CHD before surgery; 2. condition of CHD after surgery; 3. conditions of cardiomyopathies and arrhythmias, without morphological signs of CHD. The control group consisted of 20 patients, matched by sex and age. For analysis 2 ml peripheral vein blood was used in the morning.

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Results: The values of NT-proBNP, cTnT and CK were elevated in children with heart failure, early after CHD surgery, and in the active phase of cardiac arrhythmias. The highest NT-proBNP levels were found in children with HLHS and DORV. After CHD surgery, a significant drop in the value of proBNP was noticed (20.4%). The mean NT-proBNP value in children with cardiomyopathies and arrhythmias was higher than in the control group (25.2%). The highest values were recorded in patients with dilatative cardiomyopathy and SVT. In patients with PDA cardiac biomarkers level was under normal limits. Cardiac troponin T values were significantly elevated early postoperatively.
Conclusion: Cardiac biomarkers in children are valuable indicators of heart condition upon which we can assess the severity of the child's clinical condition and course of the disease, in the terms of recovery.

P2415 - OUES CUT OFF VALUES FOR CHILDREN AND ADOLECENTS COMPARING CONGENITAL HEART DISEASE VERSUS GROUP CONTROL

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Background: Cardiopulmonary testing is an important tool if evaluated by its myriad variables. Among them, the efficiency of the peripheral oxygen uptake represented by the Oxygen Capture Efficiency Scale - OUES (logarithmic regression curve between oxygen consumption and ventilation. This ratio increases in importance due to its reliability even for submaximal tests or with standards of Erratic ventilators, reflecting the muscular work, extraction and use of peripheral oxygen.
Objective: The objective of this study is to estimate cut off values for oues per kg in children healthy versus congenital heart disease and distinguish between the normal or deficit response.
Methods: Cardiopulmonary test in 305 healthy children and 371 with heart disease, divided into groups by age group. Certain OUES and other indices. The efficiency of VO2 (OUES) was analyzed for the presence or absence of cardiac change and correlated with functional capacity.
Results: The median of OUES between heart disease and the control group presented a significant difference (1217 vs 2354, p <0.05). TThe median O2 saturation at the peak of exercise was equal between the groups, but the additional finding of values lower than 70% reinforced the severity of the children with the test. The cutoff value of the OUES for the classification of subjects with heart disease was 36.6 (sensitivity 72% and specificity 62%, p <0.05). The cut of OUES for classification of individuals with normal functional capacity (>80% of predicted maximum VO2) was 35.27 (sensitivity 75% and specificity 83%, p <0.05). While the cut of OUES for individuals with values greater than 85% of the predicted maximum VO2 was 35.69 (sensitivity 78% and specificity 78%, p <0.05).
Conclusions: The results suggest that oues was able to provide an objective assessment of functional capacity in healthy and cardiac patients.

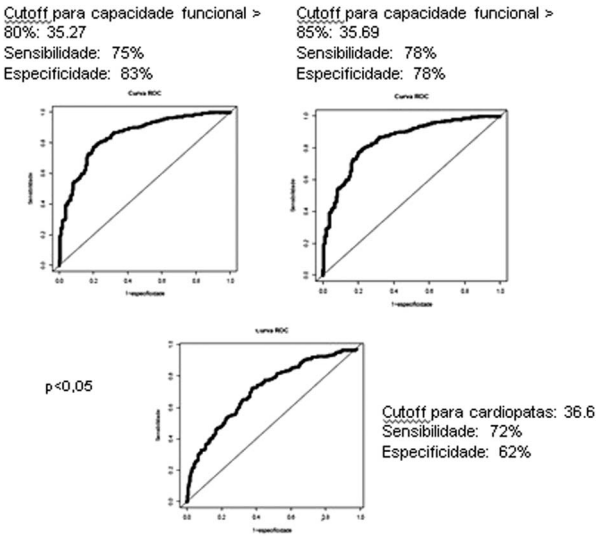

Figure 1.

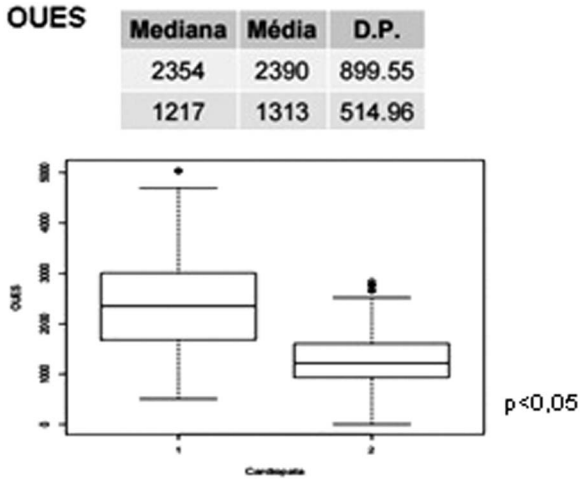

Figure 2.

P2421 - ECHOCARDIOGRAPHIC FINDINGS AND CLINICAL OUTCOME OF FETAL CARDIOMYOPATHIES

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Background: Recently, there have been few reports of fetal cardiomyopaties (CMs). However, there is a paucity of information about assessment and outcome of the disease.
Methods: We retrospectively reviewed our experience with CMs diagnosed in fetal life, from 2008 to 2016. We investigated fetal echocardiographic findings, postnatal laboratory findings, acute phase treatment course and clinical outcome.
Results: Five cases (4 boys and 1 girl) were diagnosed fetal CMs at 28 weeks to 35 weeks (median 31 weeks 6 days). All patients were diagnosed dilated cardiomyopathy. Associated left ventricular noncompaction (LVNC) were noted in 4 patients. All patients required mechanical ventilation and continued catecholamine

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infusion in the neonatal period. After postnatal treatment, all cases showed a tendency to improve symptoms of heart failure, but 3 cases worsened thereafter and died in infancy (5 months, 6 months, 9 months). Two patients survived and discharged. Cardiovascular profile score (CVPS) in fetal echocardiography was 7 to 8 points in two survivors, 4 to 6 points in three nonsurvivors. BNP levels at birth were 42 to 378 pg/ml in survivors, 1231 to 3620 pg/ml in non-survivors. LVEF was 30-35% in survivors, 18-23% in nonsurvivors. Neonatal intubation period and continuous administration period of catecholamine were shorter in survivors than in nonsurvivors (3 to 7 days vs 13 to 41 days, 7 to 10 days vs 22 to 58 days).
Conclusion: More than half cases died in infancy. Fetuses with CMs have a poor prognosis. However, it was possible that patient of higher CVPS survive and discharge after undergoing intensive care in neonatal period.

P2424 - PREDICTIVE VALUE OF CRYPTOGENIC SEROSANGUINOUS PERICARDIAL EFFUSION FOR HISTOPLASMOSIS IN AN ENDEMIC REGION

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Background: Histoplasmosis is a mycosis that is endemic to the Mississippi River valley. It's presentation ranges from asymptomatic infections to disseminated histoplasmosis. Pericarditis and pericardial effusions have been reported with varying prevalence rates; but its diagnostic value is unknown.
Methods: A retrospective review was performed at a single tertiary care children's hospital including all immunocompetent children up to 18 years of age presenting with clinically significant pericardial effusions of unknown etiology requiring pericardiocentesis or pericardial window from Oct 2012 – May 2016. The prevalence, sensitivity, specificity, and positive predictive value (PPV) of the initial presentation of histoplasmosis as a serosanguinous effusion was determined. The diagnosis of histoplasmosis was based on either positive serology or clinical diagnosis by an infectious disease specialist.
Results: There were 54, immunocompetent children that underwent therapy during the study period; of which 38 were cryptogenic. Of the 22 serosanguinous pericardial effusions, 16 were eventually diagnosed to have histoplasmosis (PPV = 72.7%). Only 4 of the 16 non-serosanguinous effusions (serous or purulent) were diagnosed as histoplasmosis. The sensitivity and specificity of serosanguinous pericardial effusion as a presentation of histoplasmosis were 80% and 66.7% respectively. The other etiologies included new onset malignancy, viral, autoimmune, or unknown. Eight of the 20 patients (40%) with histoplasmosis that presented as pericardial effusions went on to develop either pulmonary artery or pulmonary vein stenosis within 8 + 6 months; all of whose effusions were serosanguinous in character at presentation.
Conclusions: Histoplasmosis was diagnosed in a majority of immunocopetent children who presented with hemodynamically significant serosanguinous pericardial effusion of unknown etiology in an endemic region. Long term follow-up due to risk of developing additional sequelae resulting from fibrosing mediastinitis is required.

P2431 - PREEMPTIVE LIVER TRANSPLANTATION FOR HOMOZYGOUS FAMILIAL HYPERCHOLESTEROLEMIA IN CHILDREN CASE SERIES

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Background: Familial hypercholesterolemia (FH) is characterized by high serum cholesterol levels and premature atherosclerosis. Heterozygous FH common (1 in 200-500 population), but homozygous FH is rare (1 in 1 million). Normally, liver contains 50-75% of LDL receptors in the body. In homozygous FH, there is severe deficiency of LDL receptors (2-25%) causing failure to uptake LDL for metabolism and elevation of serum LDL cholesterol level in 1000 mg% range. Atherosclerosis develops early and coronary stenosis, aortic valve disease and other complications occur in second decade. Manifestations include cutaneous xanthomas, arcus in the eye, vascular plaques, atherosclerosis aortic valve disease and carotid occlusion leading to stroke. Aggressive traditional therapies (medications, plasmapheresis and bowel bypass surgeries) have been applied which only slow down the progression. Eventually, heart surgery, heart transplant and/or liver transplant is required at an early age is needed in several homozygous FH patients. Therefore, preemptive liver transplant before onset of cardiovascular manifestations of atherosclerosis has been proposed.
Methods: Retrospective review of 5 cases.
Result: Five children who showed early signs of atherosclerosis (xanthoma, carotid bruit and early plaques in carotid and or coronary arteries), failure of response to medications and underwent preemptive liver transplant were included. Preoperative total cholesterol (813-2316 mg%) and LDL cholesterol (733-1029 mg%)(Table 1). Age range at liver transplant was 2-7 years. Lipid profile normalized during immediate postoperative period and maintained during follow-up. After liver transplant, total cholesterol range was 130-201 mg% and LDL cholesterol 74-133 mg%. All patients had xanthomas which resolved after transplant (Figure 1). Follow up duration range is 3 months to 3 years. All patients were doing well at last follow-up
Conclusion: Preemptive liver transplant is an effective therapy for homozygous FH. Further follow-up is needed to establish such approach as primary therapeutic approach.

Table 1. Patient characteristics

Patient number	Preoperative Total/LDL cholesterol (mg%)	Age at liver transplant (years)	Posoperative Total/LDL cholesterol (mg%)	Length of follow-up
1	2316/733	7	161/106	2 years
2	887/772	5	183/133	3 years
3	813/791	5	130/74	1 year
4	867/804	2	154/77	6 months
5	1092/1029	3	201/114	3 months

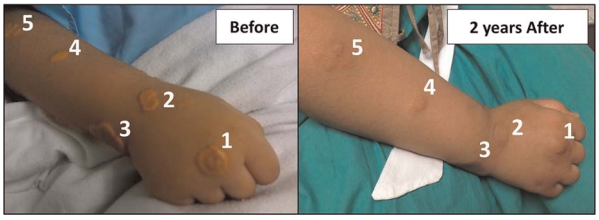

Figure 1.
Xanthomas before and after liver transplant.

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P2444 - THE NEW MODIFIED BLOOD PRESSURE TO HEIGHT RATIO SIMPLIFIES THE SCREENING OF HIGH BLOOD PRESSURE IN AMERICAN CHILDREN

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Background: High blood pressure is often underdiagnosed among children and adolescents. Many friendly tools have been described to overcome these problems. One of them, the modified blood pressure-to-height ratio (MBPHR) was tested in children between 8 and 12 years old. However, a variation of this formula (MBPHR3) showed better sensitivity and specificity in Chinese children. Therefore, confirmation on other populations is mandatory.
Material and Methods: We utilized data from the National Health and Nutrition Examination Survey (NHANES) 1999-2014. Only patients with one Systolic blood pressure (SBP) and Diastolic Blood Pressure (DBP) measurements and complete data for height, weight, gender and age between 8-12 years were included in the present study. The MBPHR were calculated as: SBP (mmHg)/ (Height(cm) + 7x(13-age(in years)) to SBP and DBP (mmHg)/(Height(cm) + 7x(13-age(in years)) to DBP. The MBPHR3 were calculated as: SBP (mmHg)/ (Height(cm) + 3x (13-age(in years)) to SBP and DBP (mmHg)/(Height(cm) + 3x (13-age(in years)) to DBP. Receiver Operator Characteristics curves (ROC) were drawn to identify optimal thresholds for each method. Afterwards, the sensitivity, specificity and positive predictive value (PPV) of each method were calculated with the respective confidence intervals. The software used was MedCalc 16.8.4 (Software bvba, Ostend, Belgium).
Results: A total of 6587 cases were analyzed. Exactly 3.92% of the participants were pre-hypertensive and 2.84% hypertensive. MBPHR3 showed higher specificity and PPV than MBPHR and BPHT, with similar sensitivity, for pre-hypertension or hypertension (MBPHR3: 93.12%, 92.66% and 99.47% for males and 96.04%, 89.94% and 40.98% for females. MBPHR: 96.79%, 86.23% and 33.44% for males and 95.15%, 90.33% and 41.94% for females).
Conclusion: MBPHR3 showed better results in American children aged 8-12 years with a low number of cut-off points.

P2447 - GENDER DIFFERENCE IN THE PREVALENCE OF CONGENITAL HEART DISEASE IN DOWN'S SYNDROME A BRIEF META ANALYSIS

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Background: Down's syndrome (DS) affects one per 700 live births and congenital heart disease (CHD) occurs in 40-60% of these patients. Many factors contributes to the origin of CHD in DS and gender could be one of them. The purpose of this study was to compare the prevalence of CHD and DS between genders through a meta-analysis and systematic review.
Material and Methods: Studies that described the prevalence of CHD in DS and included gender information for each defect were included. The search for articles was performed in the following data sources: Medline (accessed via Pubmed), Scopus and Scielo. Terms utilized were those used by Mesh for Medline and Scopus, and the descriptors of Health Sciences (Decs) for Scielo. The terms included: "prevalence", "Down syndrome" and "congenital heart disease". A combined data analysis was performed to identify the Odds Ratio between genders, being the female gender considered a risk factor. The confidence intervals and the size of the pondered effect were calculated and the meta-analysis graphs built using the MedCalc v 16.8 software. Random effects model was utilized.
Results: 595 abstracts were selected for analysis, but only four filled the eligibility criteria and were included in the meta-analysis totaling 8580 patients with DS (4726 from the male gender and 3854 from the female gender). It is observed that female gender is a risk factor for the presence of CHD in Down's syndrome (OR: 1.398; 1.143 to 1.709). The same occurs if we consider AVSD alone (OR: 1.421; CI: 1,252 to 1.613). However, when VSD and ASD are considered, there is no difference among groups.
Conclusion: this meta-analysis showed a clear trend towards a higher prevalence of CHD in the female population with Down syndrome.

P2448 - COMPARISON OF CAPTOPRIL AND VALSARTAN IN CHILDREN L TO R SHUNT WITH HEART FAILURE

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Background: In children, heart failure is most often caused by L to R shunt which the management has largely evolved based on clinical experience and the application of adult data, supported by the more limited paediatric literature. Valsartan are increasingly used in heart failure in adults, especially where there is intolerance to captopril. There is no published evidence supporting their use in children with the condition in Indonesia.
Objectives: The aim of this study was to compare the effects of captopril and valsartan in children L to R shunt with heart failure.
Methods: Randomized controlled trial was conducted on 1-16 year-old L to R shunt with heart failure.Thirty two patients were randomized into 2 groups to received captopril either valsartan. We performed examinations including PHFS, echocardiography, electrocardiography, and chest x-ray before and 4 weeks after treatment. Both group had also received diuretic spironolacton. Statistical analysis was carried out using paired t-test, t-test, Mann-Whitney test, and independent t-test.
Results: Treatment with captopril resulted in significant improvement of PHFS from 7.06 ± 2.04 to 4.75 ± 2.43 ($p < 0.0001$; 95% CI -2.98 to -1.65); reduction of heart rate from $117,75 \pm 14,67$ times/minute to $109,63 \pm 17,59$ times/minute ($p < 0.05$; 95% CI -15.78 to -0.46). While valsartan resulted in significant improvement of PHFS from $6,81 \pm 2,25$ to 3.94 ± 1.98 ($p < 0.0001$; 95% CI -3.76 to -1.98); reduction of heart rate from 117.1 ± 21.86 times/minute to 108.6 ± 20.66 times/minute ($p < 0.05$; 95% CI -14.17 to -2.83), decrement of S Amplitude in lead V1 from 10.06 ± 6.78 to 7.31 ± 5.99 mm ($p < 0.05$; 95% CI -4.69 to -0.81), decrement of CTR from 59.74 ± 4.72% to 57.19 ± 5.14% ($p < 0.001$; 95% CI -3.87 to -1.23). In our study, RR 0.85, ARR

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0.125, RRR 0.143. Valsartan have lower rates of cough than captopril (NNH = 8).
Conclusion: Valsartan had better effect than captopril and could be one of choice beside captopril in treating heart failure.

P2450 - NEW EQUATIONS CAN IMPROVE THE DIAGNOSIS OF HIGH BLOOD PRESSURE

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Background: high blood pressure is usually underdiagnosed in children and adolescents. One of the factors that determine this is the complicated diagnosis process. Several screening methods were developed to circumvent those problems, and each has advantages and disadvantages. A method that combines simplicity and the non-use of additional tools can change this context.
Methods: linear regressions correlating height with the 90th percentile of systolic and diastolic blood pressure for each year of age and sex were calculated (?). The values of the constants obtained were rounded to the closer integer value to make it easier to remember. Then, the equations were tested and the sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) for high blood pressure identification were calculated. We utilized data from the National Health and Nutrition Examination Survey (NHANES) 1999-2014. Only patients with one Systolic blood pressure (SBP) and Diastolic Blood Pressure (DBP) measurements and complete data for height, weight, gender and age between 8-17 years were included in the present study. A total of 14167 patients was analyzed. The utilized software was MedCalc 16.8.4 (Software bvba, Ostend, Belgium).
Results: the obtained formulae were: 70 + height/3 for SBP and 40 + height/4 for DBP for both genders. Formulae presented high sensitivity (92.75%; CI: 90,97% to 94,28%), specificity (98,66%; CI: 98,45% to 98,85%), NPV (99,44%; CI: 99,30% to 99,56%) and PPV (84,14%; CI: 81,86% to 86,25%). The frequency of pre-hypertension and hypertension in this population was 7.1%.
Conclusion: the use of the described equations can aid in the identification of children and adolescents with high blood pressure. Among its advantages are the possibility of its use in different populations, the need of no additional tools and accuracy of results for both genders.

P2455 - POTASSIUM INTAKE AND HIGH BLOOD PRESSURE IN CHILDHOOD

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Background: Hypertension is a worldwide public health problem, because of its intimal correlation with conditions of high mortality like stroke. Its prevalence is increasing among children, as a consequence of the obesity epidemics. Additionally, different patterns of consumption of micronutrients can lead to or protect from hypertension. This study aimed to compare the food intake of sodium and potassium between children with and without high blood pressure.
Material and Methods: parents of children participating in an active search for congenital heart disease in 13 different cities from Paraíba – a state form northeast Brazil – in July 2016 were included in this study. They answered a food intake questionnaire for their children, from the previous day. Then, the intake of sodium and potassium were calculated from tables of Instituto Brasileiro de Geografia e Estatística (Brazilian Institute of Geography and Statistics). Non-parametric tests were performed to search for correlation between the presence of high blood pressure and the intake of such nutrients. Only preschool and school children were included.
Results: A total of 225 children (54.12% male) were analyzed. Exactly 115 were preschool children and 47 had obesity. According to the measured blood pressure, 20 children had high blood pressure (prehypertension or hypertension). A significant correlation between obesity and high blood pressure was encountered ($p < 0.01$). The intake of potassium was higher in children without high blood pressure ($p = 0.0254$), but no correlation was seen between sodium intake and high blood pressure ($p = 0.7762$). A lower sodium to potassium ratio correlated with normal blood pressure ($p = 0.4488$).
Conclusion: a poor potassium intake from childhood may lead to high blood pressure. Nutrition counseling from early ages may contribute not only to the health of the child but throughout an individual's life course.

P2462 - PHYSICAL ACTIVITY MODULATES ARTERIAL STIFFNESS IN CHILDREN WITH CONGENITAL HEART DISESASE

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Background & Hypothesis: Children with congenital heart disease (CHD) are often less physically active compared to healthy children. Children with CHD are also at risk for developing greater arterial stiffness and physical inactivity can potentiate this risk. We tested the hypothesis that less physically active children with CHD would have greater arterial stiffness compared to more physically children with CHD.
Materials & Methods: Nineteen children with CHD (11 ± 3 years; males = 9) and 20 age- and sex-matched controls (11 ± 3 years; males = 11) were studied. Carotid-radial pulse-wave velocity (PWV) was assessed with applanation tonometry to determine arterial stiffness. A 6-minute walk test (6MWT) was performed to estimate aerobic fitness. Average daily step count was assessed by accelerometry over a 7-day period. The median step count score for each group was used to determine high-step (high physical activity) and low-step (low physical activity) groups. Data were analyzed with t-tests and significance accepted at $P < 0.05$.

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Results: 6MWT distance was lower in CHD (521 ± 50 vs. 605 ± 79 m; $P < 0.001$). PWV was similar between CHD (8.78 ± 1.24 m/s) and controls (8.67 ± 1.28 m/s; $P > 0.05$). However, when considering physical activity as a modulating factor for PWV (by comparing high-step and low-step groups), PWV was significantly lower in the high-step CHD group (7.89 ± 0.68 m/s) compared to the low-step CHD group (9.78 ± 0.91 m/s; $P < 0.001$), but not between the high-step (8.70 ± 1.35 m/s) and low-step control group (8.64 ± 1.28 m/s; $P > 0.05$).
Conclusion: Physically activity in children with CHD may be an especially important modulating factor for arterial stiffness.

P2464 - PRESENCE OF CONGENITAL HEART DISEASE IN NEONATES WITH AND WITHOUT MICROCEPHALY AFTER ZIKA OUTBREAK A COMPARATIVE STUDY IN NORTHEAST BRAZIL

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Background: Brazil faced an increase in number of microcephaly cases last year. Until now, the ZIKA virus is being considered the main responsible for the phenomenon. Besides microcephaly, other malformations can occurred in infected neonates. Congenital heart disease (CHD) could be one of them. Therefore, the objective of this work was to compare the presence of CHD between neonates with and without microcephaly after ZIKA outbreak.
Material and Methods: Comparative study, based on a retrospective data analysis. The data were obtained from Rede de Cardiologia Pediátrica Pernambuco-Paraíba (Pediatric Cardiology network Pernambuco-Paraíba). The Network uses pulse oximetry and screening echocardiograms to diagnosis neonates with CHD. Neonates born after the first semester of 2014 with informed head circumference, gestational age above 37 weeks and echocardiographic results were included. A head circumference smaller than 32 cm was considered microcephaly. The chi-square test was utilized to compare categorical variables. $P < 0.05$ were considered significant.
Results: A total of 788 patients were analyzed. Only 39 (4.9%) had microcephaly. In relation to gender, 438 were female (55.6%). CHD was diagnosed in 334 cases. No statistical correlation between microcephaly and CHD was observed ($p = 0.8601$). The main diagnoses were septal defects (223), patent ductus arteriosus (199), and pulmonary stenosis (25). Complex lesions were present in 16 had complex lesions. No statistical significance was obtained between each type of CHD and the presence of microcephaly.
Conclusion: There seems to be no relation between microcephaly and CHD. However, it is still necessary to exclude the association between microcephaly and functional anomalies or arrhythmias.

P2480 - RISK OF RESPIRATORY SYNCYTIAL VIRUS INFECTION IN CONGENITAL HEART DISEASE IN A TROPICAL COUNTRY

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Background: The impact of respiratory syncytial virus (RSV) infection in infants and children with congenital heart disease is relatively understudied in Southeast Asian tropical countries. In temperate countries, seasonal activity of RSV has been reported, particularly in association with temperature, while variable trend of RSV activity has been found in tropical countries.
Objective: To compare the risk of RSV-associated hospitalization and analyze the epidemiology of RSV infection in patients with cyanotic and acyanotic congenital heart disease (CHD) in tropical Singapore, where RSV immunoprophylaxis is not routine practice.
Method: This was a single-centre, retrospective study, including patients diagnosed with CHD from 2005-2010. The study cohort included 75 patients with cyanotic CHD and 225 patients with acyanotic CHD. Patients with acyanotic CHD were further classified into hemodynamically significant (hs)-acyanotic and non-hs-acyanotic groups according to whether they underwent surgery or took at least 2 anticongestive medications.
Results: RSV-associated hospitalization rate was similar in all 3 groups. However, intensive care admission (15% vs 9% vs 8%, $p = 0.01$) and high-dependency admission rates (23% vs 19% vs 4%, $p = 0.03$) were higher in the cyanotic group than in the other 2 groups. Logistic regression revealed that cyanotic CHD was the most significant risk factor for the ventilator support and RSV-associated morbidities. In both patients with cyanotic and acyanotic CHD, RSV-associated hospitalization rate was higher in patients aged younger than 1 year, and in the middle of the year (May – Aug) in Singapore, a tropical country.
Conclusions: The results show that patients with cyanotic CHD have a higher risk of severe RSV infection than do those with acyanotic CHD. RSV prophylaxis is more important and may reduce costs more for patients with cyanotic CHD.

P2486 - THE IMPROVEMENT CLINICAL MANIFESTATION AND CHEST X RAY AFTER CARVEDILOL THERAPHY IN HEART FAILURE DUE TO CONGENITAL HEART DISEASE LEFT TO RIGHT SHUNT

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Background: Heart failure due to congenital heart defect (CHD) left to right shunt are increased pulmonary blood flow and caused volume overload. Conventional therapy has not blocked the sympathetic system yet. Carvedilol, a novel non selective β-blocker, reduces mortality and hospitalization in adults with heart failure. Limited information is available about its use in children.
Objective: To evaluate the effect of carvedilol for changes of pulse rate, respiratory rate, hepatomegaly, and chest x-ray in children with heart failure due to CHD left to right shunt.
Methods: A randomized, double-blind, placebo-controlled study was done. In addition to conventional therapy (ACE inhibitors and diuretics), patients were assigned to receive placebo or carvedilol. Carvedilol was initiated at a dose of 0.05 mg/kg/day, with a target dose of 0.2 mg/kg/day. The outcomes of pulse rate, respiratory rate, and hepatomegaly presence were evaluated before-after treatment. Chest x-ray was done before-after treatment and evaluating for cardio-thoracic ratio (CTR) and pulmonary vascularity. The data was analyzed using independent sample t-test and Chi-square test, with confidence interval 95%.

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Results: Of 30 patients, 15 in each groups. The mean age was 57.6 (SD 43.57) months, 19(63.3%)were boys. There were 21(70%) children with VSD and 9(30%) children with PDA. Compared to control group, there were greater reductions of pulse rate (-19.2 ± 7.5) and respiratory rate (-7.2 ± .8) in carvedilol group with p value <0.0001. No hepatomegaly. The carvedilol group had a significant decreasing of CTR(-2.94 ± 2.34% versus -0.48 ± 3.19%, p = 0.023, CI 95%:-4.556 to-0.360). However, there was no significant change of pulmonary vascularity (p = 0.153).
Conclusion: Carvedilol decreases the pulse rate, respiratory rate, and the CTR on chest x-ray, but does not decrease hepatomegaly, and the pulmonary vascularity.

P2492 - THE INCIDENCE AND OUTCOMES OF ASYMPTOMATIC VENTRICULAR SETPAL DEFECTS DETECTED BY NEONATAL SCREENING ECHOCARDIOGRAPHY

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Background: Ventricular septal defect is the most common type of congenital heart disease. Echocardiography can easily provide an accurate diagnosis of these cardiac defect.This study focuses on the incidence & short-term outcomes of VSD which were diagnosed by using screening echocardiography in asymptomatic neonates.
Methods: From January 2009 to June 2015, a total of 10150 full-term newborn babies were delivered in our hospital. Of them, 3125 received screening echocardiographic study. The echocardiography was performed within 3 days of life by two well-trained technicians. Besides the other 135 neonates with complex CHD, the various subtypes of simple VSD were picked up on 61 neonates (1.95%). Forty-five neonates who had complete perinatal data and regular follow-up studies were enrolled for analysis. The gender, birth body weight, parity, family history of CHD, methods of delivery, & subtypes of VSD were analyzed by using statistical software SPSS. The spontaneous closure rate & outcomes of various subtypes of VSD were also analyzed
Results: Of the 45 neonates with VSDs, there were subpulmonic in 1 case, perimembranous type in 14 cases, atrioventricular type in 0 cases & muscular type in 30 cases. The p valve of relationship between the occurrence of VSD with the gender, birth body weight , family history of CHD, and method of delivery are 0.745,0.763,0.872 and 0.436 respectively. Spontaneous closure of VSD was noted 72.41% in perimembanous type 43.48% and 86.49% in muscular type during the 3 months to 6 years (7.28months ± 5.18 months) follow-up There are no patients developed the symptoms and signs of congestive heart failure needed medical therapy. 2 patients underwent surgical closure . 31 cases are followed-up in out-patients clinic regularly.In addition, the p value the spontaneous closure is 0.041(p < 0.05).
Conclusions: The incidence of VSD in asymptomatic neonates is 1.95% which is much higher

P2501 - CONGENITAL UNILATERAL ABSENCE OF PULMONARY ARTERY SINGLE INSTITUTIONAL EXPERIENCE IN A LOW MEDIUM INCOME COUNTRY

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Background: Unilateral absence of pulmonary artery (UAPA) is a rare congenital cardiac malformation characterized by complete absence of the intrapericardial segment of one of the main pulmonary arteries.It can present as an isolated lesion (40%) or may be associated with other congenital heart defects (CHD) in approximately 60%. Isolated UAPA is associated with increased morbidity and mortality related to pulmonary hypertension (PHT) and recurrent haemoptysis. Surgery is limited to the intracardiac anomalies if no identifiable PA in the hilum.
Methods: Retrospective review of 35 cases of UAPA seen at the Chris Hani Baragwanath Academic Hospital, a tertiary care institution between 1980-2016.
Results: Twenty infants(57,1%) and fifteen children were reviewed.Median age at presentation was 10 months (1day-11years). UAPA was isolated in 15 patients(42,9%) and associated with CHD in 20 patients(57,1%). Thirty three patients (94,3%) were symptomatic. Common presenting feature was respiratory distress in isolated UAPA(73.3%) and cyanosis in associated UAPA (47.9%). Chest X-ray was suggestive of UAPA in all patients with isolated UAPA and nine patients(45%) with associated UAPA. The diagnosis of UAPA was confirmed on computerized tomography (CT) scan, angiography and at surgery in thirty one patients(88.6%). Unilateral absence of right pulmonary artery (UARPA) was identified in 80% of the isolated UAPA as compared to the UALPA identified in 75% OF associated UAPA. Pulmnary hypertensio(PHT) was common in patients with isolated UAPA(53.3%) and the majority were infants(75%). Collateralls were identified on angiography in twelve patients (34,3%). Congenital heart defects with UAPA were as follows: Ventricular septal defect(VSD) = 3; Patent ductus arteriosus (PDA) = 3; Common atrium = 1;Double inlet left ventricle (DILV) = 1; Tetralogy of Fallot(TOF) = 9; Double outlet right ventricle(DORV) with pulmonary stenosis(PS) = 3. Surgical anastomosis was performed in one patient with TOF associated with UALPA.Surgery for other associated congenital cardiac defects was performed in eighteen patients(51.4%). Overall mortality was 34,3%.
Conclusion: Mortality is high in association with UAPA. Isolated UAPA and associated UAPA with increased PBF (pulmonary blood flow) were associated with PHT. Isolated UAPA was commonly associated with UARPA while UALPA was found in majority of associated UAPA.

P2539 - ARTERIAL ISCHEMIC STROKE IN CHILDREN WITH CONGENITAL HEART DEFECT

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Introduction and Objectives: A third of paediatric arterial ischemic stroke (AIS) is related to procedures in patients with congenital heart defects (CHD). An incidence of AIS with motor deficit is estimated in 2-6% of these patients, with higher rates in surgeries requiring extracorporeal circulation. Our objective is to describe the characteristics of AIS in children with CHD.
Methods: retrospective descriptive study of patients with CHD who suffered an AIS followed in 2 visits to outpatient clinics of

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Child Cardiology during 8 years. We analysed: heart disease, previous procedures, clotting test, clinical and diagnostic delay, prophylaxis, and neurological sequelae.
Results: 10 cases of AIS in CHD were collected (see chart below). In all cases, the neurological disease was noticed in the first 6 months after surgery, except in one case that was delayed up to 5 years. In 4 patients, the procedure prior to AIS was a catheterization, in the other 6 was major surgery, two of them with postoperative extracorporeal membrane oxygenation (ECMO). Clotting test was performed in 6 of the cases, being positive for the same mutation in two of them, not related. The neurological consequences were in 7 of the cases cerebral palsy(CP) type Spastic Hemiparesis (SH), that one of the cases was transient. Intellectual disability occurs in 5 of our patients, in one case in isolation, and in others, associated with focal epilepsy or developmental delay. Two patients are under tests due to suspicion of metabolic pathology.
Conclusions: In a child with CHD, it is a priority to think about AIS in the presence of any neurological symptom, especially focal, and if there is a history of recent invasive procedures. In these cases, clotting tests should be considered as a cause of comorbidity. a multidisciplinary management of these patients is basic in order to minimize the sequelae.

Table.

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7	Patient 8	Patient 9	Patient 10
Age at diagnosis of AIS	10 m.o.	12 m.o.	6 m.o.	5 y.o.	2 y.o.	1 m.o.	7 m.o.	26 m.o.	3 m.o.	8 m.o.
CHD type	VSD	VSD + pulmonary atresia	VSD + pulmonary stenosis	ToF	Hypoplastic left heart syndrome	CoA & Taussig–Bing syndrome	Scimitar syndrome and pulmonary hypoplasia	ASD & Primary Pulmonay Hypertension	ToF	Pulmonary atresia
Procedure prior to AIS	Major surgery	Catheterization	Major surgery	Major surgery + ECMO	Catheterization	Major surgery + ECMO	Major surgery	Catheterization	Major surgery + ECMO+ sepsis+ endocarditis	Catheterization
Genetic Clotting test	Normal	F2 20210G>A mutation	Normal	Normal	No	No	No	F220210G>A mutation	Normal	No
Primary prophylaxis	Antiaggregation	No	Antiaggregation	No	Anticoagulation	No	No	No	No	Anticoagulation
Motor focal manifestations	CP, left SH	Transient left SH	CP, right SH	Left SH	CP, left SH	No	No	Left SH	No	CP, Right SH
Other neurological manifestations	Focal epilepsy	No	No	No	Intellectual disability	Focal epilepsy, Mild developmental retardation.	Intellectual disability	Intellectual disability	Developmental retardation.	No

VSD: Ventricular septal defect; ToF: tetralogy of Fallot; CoA: Coarctation of the aorta; ASD: atrial septal defect.

P2541 - DIAGNOSES AND OUTCOMES OF THE PATIENTS EVALUATED IN A CONSULTATION OF FETAL ECHOCARDIOGRAPHY IN THE PAST 10 YEARS

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Introduction and Objectives: Congenital heart disease affects approximately 1% of live births. Our objective was to describe the clinical characteristics and the fetal evolution of pregnant women referred to the Perinatal Cardiology Clinic in our hospital.
Material and Methods: A descriptive, retrospective study that included the pregnant women referred to the Perinatal Cardiology Clinic at our hospital from its implementation in 2006 until December 2015.
Results: A total of 481 ultrasound scans where performed in 220 different gestations, with an average of 2.18 ultrasounds per gestation. Pregnant women from several provinces of the Community were seen. 35% of gestations were in the second trimester, 65% in the third. 47% were referred for suspicion of structural heart disease, 7% for suspected arrhythmia, 3% for anomalous cardiac echogenicity, 33% for family history of heart disease, 3% for maternal disease and 5.4% for pregnant women without risk factors The suspicion of structural heart disease was confirmed in 64.7% of fetuses, with ventricular septal defects being the most frequent pathology (32%), followed by aortic anomalies (8.8%), tetralogy of Fallot and transposition of large arteries . After the diagnosis of heart disease, 22 patients voluntarily terminated their pregnancy and 47 pregnancies followed their natural evolution, with an intrauterine death fetus at 38w of gestacional age. Of the complex heart diseases, one patient required heart transplantation at birth, the others received corrective surgery with good evolution, except for 4 deaths in the first two years of life due to complications of their heart disease.
Conclusions: Fetal echocardiography is an essential tool in the diagnosis of congenital heart disease, which allows planning the right place and time for delivery and provides an early treatment for newborn affection, especially relevant in communities without Congenital Heart Surgery such as ours.

P2545 - ADOLESCENTS IN THE CARDIOLOGY CONSULTATION

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Introduction and Objectives: The reason for consultation in Pediatric Cardiology varies according to age. The objective of the study was to describe the characteristics of the consultations performed in the Cardiology Clinic by adolescents.
Material and Methods: A retrospective descriptive study of the clinical evolution of patients between 10 and 14 years of age, derived from our Clinic, in a 1 year period.
Results: 134 adolescents came to a first consultation inthis period. The causes of referrals where: 3% family history of heart disease, 5% personal cardiac history, 10% suspected arrhythmia on routine electrocardiogram, 12% cardiologic study due to another disease, 15% murmur in clinical examination and 55% presence of Cardiovascular symptoms (35% palpitations, 23% chest pain, 27% presyncope and 15% dyspnea). All patients with a personal history had normal complementary tests. Only one case with family history presented positive tests. 3 of the patients with systemic pathology presented cardiac involvement. Among patients who consulted because of a murmur, 2 coronary fistulas and 4 minor structural heart defects were diagnosed. 8 arrhythmias were confirmed, 2 of them where severe. Regarding symptom consultations, palpitations were equally present in both sexes. Presyncope was more frequent in women, and only one of them presented complementary positive tests. Chest pain did not show differences between sexes, and only one case showed cardiac disease . Dyspnea was more frequent in men with 78% of them showing abnormal spirometries.
Conclusions: In our study, the most frequent cause of consultation in adolescent patients was the presence of cardiovascular symptomatology, which causes great distress in the patients and their families, being the complementary tests in the majority of them negative, with the exception of the ones who presented alarm symptoms in the anamnesis.

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P2547 - AORTIC STIFFNESS ECHOCARDIOGRAPHIC ASSESSMENT IN DIABETES MELLITUS TYPE 1 PEDIATRIC PATIENTS

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Introduction: Among vascular complications of diabetes mellitus, pediatric arterial stiffness has shown to be a cardiovascular disease predicting factor in adult age. Our objective was to use echocardiography for the assessment of aortic stiffness in a group of patients and its association with metabolic control and physical activity.
Material and Methods: Case-control descriptive cross-sectional study. 44 cases: DM1, under 18 years old and without cardiovascular symptoms. 51 controls, healthy, with same age and anthropometric measurements. Aortic function was assessed calculating aortic diameter change (mm) = aortic systolic diameter (SD)– aortic diastolic diameter (DD). Aortic strain = (SD-DD)/DD. Elastic modulus E(p) = (Systolic Blood Pressure (SBP) – Diastolic Blood Pressure (DBP))/Aortic tension. Aortic Stiffness (βindex) = In (SBP/DBP). Aortic Distensibility = (2 x strain)/(SBP-DBP).
Results and Discussion: Healthy patients showed a more elastic aorta (t(48): -1.748, $p < 0,05$) than cases (t(38): -1.748, $p < 0,05$).

- DM1 patients had greater aortic stiffness while decreased distensibility, elasticity and strain than healthy controls.
- Aortic systolic and diastolic diameters and aortic velocity were similar in both cases and controls.
Linear Regression:
Aortic Distensibility: -0.004 + 0.05. weekly METs -0.001. HbA1C – 0.005. Non HDL cholesterol -0.001 current age.
Aortic Strain: 0.06-0.001. Non HDL cholesterol +0.001 weekly METs.
Our work, as found in available bibliography, but assessed by a simpler technique, concluded that DM1 patients have significantly decreased elasticity and distensibility indexes while greater aortic stiffness, even in preadolescent patients

P2550 - CARDIOVASCULAR FUNCTION EVALUATION BY ECHOCARDIOGRAPHY IN DIABETES MELLITUS TYPE 1 PEDIATRIC PATIENTS

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Introduction: Vascular complications are the most important cause of morbidity and mortality in Type 1 diabetes mellitus (DM1) patients. Our objective was to use echocardiography to evaluate cardiovascular repercussions in these patients and its association with physical activity and metabolic control.
Material and Methods: Case-control descriptive cross-sectional study. 44 cases: DM1, under 18 years old and without cardiovascular symptoms. 51 controls, healthy, with comparable anthropometric features and age. MAPSE, E' and S' waves were used to measure systolic function; TAPSE, E/A ratio, insovolumetric relaxation time (IRT), E'/A' and E/E' ratio for diastolic function and Tei index for global function.
Results and Discussion:
SYSTOLIC FUNCTION:
- A higher E/E' ratio was found in diabetic patients, a persistent difference in the evaluated groups.
- E/A, E'/A', TAPSE and IRT were similar in both cases and controls. DIASTOLIC FUNCTION: Healthy, sports inclined patients had higher S' waves than sedentary ones. - E' and MAPSE were comparable in all groups. - S' was similar in both groups. Tie Index: Results were higher in cases than in controls, but similar between subgroups. LINEAL REGRESSION: E/E' ratio: 4.194 + 0.23. Non HDL cholesterol – 0,044. Weekly METs. Our findings concur with previous bibliography on the subject, for we found diastolic dysfunction with normal systolic function in pediatric and preadolescent diabetic patients. We found E/E' ratio to be the most useful for diastolic function evaluation, showing in addition a direct association with non HDL cholesterol levels and indirectly with active lifestyle. Tei index, which evaluates global cardiovascular function, was higher in cases than in controls, probably at the expense of diastolic function. Furthermore, we found an association between this index and HbA1C levels.

P2552 - ELECTROCARDIOGRAPHY APPLICABILITY ON CARDIOVASCULAR EVALUATION OF DIABETES MELLITUS TYPE 1 PEDIATRIC PATIENTS

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Introduction: Type 1 diabetes mellitus (DM1) is one of the most common chronic diseases in infants, having great public health impact globally nowadays. Besides acute complications, vascular complications are the most significant cause of morbidity and mortality in these patients. Routine follow-up has included microvascular diseases screening for a long time. Our objective was to evaluate cardiovascular status in DM1 pediatric patients using electrocardiography.
Material and Methods: Case-control descriptive cross-sectional study. 44 cases: DM1, under 18 years old and without cardiovascular symptoms. 51 controls, healthy, with similar age and anthropometric characteristics.
Results and Discussion: Regarding electrocardiographic intervals, the only statistically significant difference ($p < 00,5$) were shorter ST intervals found in DM1 patients, compared with controls. QRS and P waves were similar in both groups. T wave axis did show a statistically significant difference ($p < 00,5$), being smaller in DM1 cases; it was between 0° and 90° in all subjects. QRS-T didn't display any significant differences. Sokolow index measurement revealed statistically significant differences ($p < 00,5$) among both groups, with only one DM1 patient in pathological range. The previously described statistically significant differences found in the electrocardiographic records have no clinical significance, therefore we do not consider this an appropriate cardiovascular screening technique in DM1 patients.

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P2585 - DIAGNOSIS AND MANAGEMENT OF CONGENITAL RIGHT CORONARY ARTERY ANOMALIES A SINGLE CENTER EXPERIENCE

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Background: Anomalous origin of the right coronary artery (ARCA) is a rare congenital heart disease that may cause sudden death. Since most patients are asymptomatic and its detailed natural history remains obscure, diagnosis is challenging and clinical management is controversial.
Objective: To investigate clinical course and short-term prognosis of ARCA cases at a single center covering approximately 70,000 children population during the last 10 years.
Design/Methods: Patients referred to Nagasaki University Hospital between 2005 and 2015 were recruited and underwent echocardiography, multi-detector computed tomography (MDCT), exercise ECG and myocardial scintigraphy. Medical charts of ARCA cases without any other cardiovascular complication were reviewed retrospectively for clues to make diagnosis, athletic activities, symptoms, evidence of ischemic condition, and therapeutic interventions.
Results: Six ARCA cases (4 males and 2 females), aged 6-17 years, were identified and followed up for 4-15 years. They were demonstrated by echocardiography and confirmed by MDCT. Two were siblings. Three were referred for chest pain. Two were referred for a complete right bundle branch block on a regular ECG examination at school. Although minimal abnormal findings were detected by exercise ECG in one and by myocardial scintigraphy in two, they were transient and poorly reproducible. All patients are permitted to take moderate exercise, if they feel no chest discomfort. None of them underwent surgical procedures since there was no apparent evidence of ischemia. None has developed myocardial infarct or died so far.
Conclusions: Siblings need the screening, since there sometimes exist familial cases of ARCA. Chest pain provided diagnostic clues most commonly in this study; however, neither exercise ECG nor myocardial scintigraphy evidently showed ischemic change. Although short-term prognosis appears good, it is difficult to determine whether surgical intervention is required for ARCA cases without apparent ischemic change. Further studies are needed to set up better clinical management.

P2595 - PREVENTION OF ADULT CARDIOVASCULAR DISEASES IN EMIRATI CHILDREN – A PRELIMINARY REPORT

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Background/Hypothesis: Atherosclerosis starts early in life and remains asymptomatic in children. Its progression is hastened by obesity, hypertension, diabetes, and dyslipidemia. Early recognition and prompt intervention of these adverse events would halt the development of cardiovascular diseases (CVD) in adults.
Aims: (1) Determine the prevalence of obesity and hypertension in Emirati children; and (2) Engage students with increased susceptibility to CVD in risk-reduction and counselling programs.
Methods: Study subjects were children in grade 2 (elementary school), 6 (middle school) and 10 (high school). Parents completed the study questionnaire, which addressed consanguinity and CVD risk factors. Students had height, weight, body-mass-index (BMI), waist circumference and blood pressure (BP) determinations. The International Obesity Task Force (IOTEF) criteria were used for BMI classification.
Results: Five-hundred-thirty-five students were included in this study; their mean [SD] age was 10.7 [11] years (M:F = 1:1). Risk factors identified in their fathers were smoking (21%), hypertension (14%), dyslipidemia (12%), heart attacks (3%), diabetes (16%), and obesity (9%). Risk factors identified in their mothers were hypertension (4%), dyslipidemia (5%), diabetes (6%), and obesity (3%). The prevalence of consanguinity was 38%. For male students, the prevalence of thinness was 31%, overweight 14%, obesity 6%, and extremely-obesity 3%. For female students, the prevalence of thinness was 40%, overweight 9%, obesity 5%, and extremely-obesity 3% (Fig. 1). The z-scores of systolic and diastolic BP increased with increased excess body fat (Fig. 2). Fig. 3 represents the waist circumference percentiles in function of age.
Conclusions: Hypertension and obesity were common among the studied children. Affected students require counseling and effective interventions.

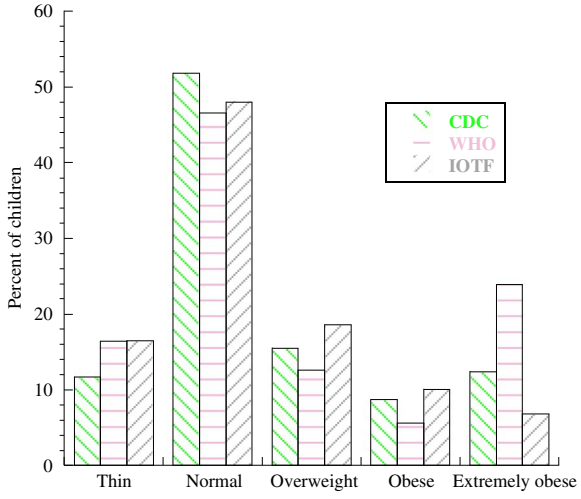

Figure 1.
IOTEF classification of the BMI of the studied students.

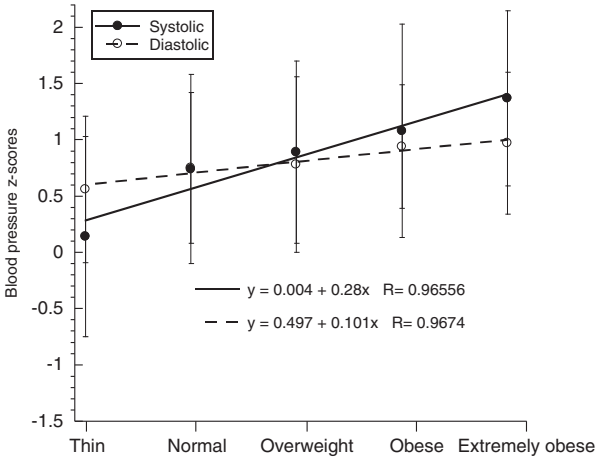

Figure 2.
Systolic and Diastolic BP z-scores in relation to excess body fat.

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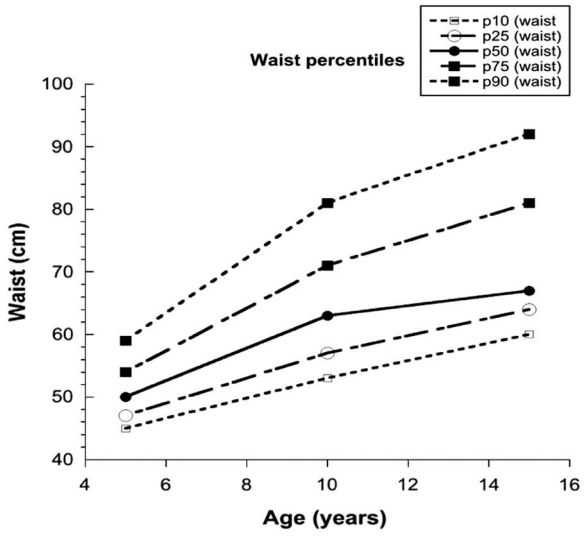

Figure 3.
Waist circumference percentiles in function of age.

P2599 - DEVELOPMENT AND VALIDATION OF THE BRIEF DEVELOPMENTAL ASSESSMENT IN PRE SCHOOL CHILDREN WITH HEART DISEASE

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Background: Despite awareness of neurodevelopmental abnormalities in children with congenital heart disease (CHD), there is no routine monitoring in the United Kingdom, with a potential for delayed detection and intervention. To address this, we developed an easy to administer early recognition tool – the Brief Developmental Assessment (BDA) - targeted for children with CHD. The BDA covers 5 age bands (0-4 months, 5-8 months, 9-14 months, 15 months-2.9 years, 3-4.9 years), and allows categorisation as green (appropriate for age), red (delayed) or amber (equivocal), can be administered by non-developmental specialists, and without specialised equipment.
Methods: A convenience sample of 960 pre-school children with CHD from each of the five age bands was recruited from three UK tertiary cardiac centres. The BDA was prospectively validated using Mullen Scales of Early Learning (MSEL). Internal reliability was explored using Cronbach's alpha. Construct validity was based on BDA detection of children with known neurodevelopmental abnormalities, and sensitivity and specificity of BDA was evaluated against MSEL.
Results: The BDA was successfully validated in the older four age bands (not those <4 months) as pre-set validation thresholds were met (lower 95% confidence limit for the correlation coefficient above 0.75) between two raters and with MSEL. Based on American Association of Pediatrics Guidelines, which state that the sensitivity and specificity of a developmental screening tool should fall between 70 and 80%, the BDA outcome of 'red' met threshold for detection of MSEL scores >2 standard deviations below the mean.
Conclusions: The BDA, along with a user-training package and an action guide, may be used to improve the quality of neurodevelopmental assessment of children with CHD, facilitating early detection and increased access to services. Further research is needed to determine the best approach in infants <4 months of age and assess progression over time in children with CHD.

P2605 - LOW INCIDENCE OF "TYPICAL" ECG FINDINGS IN 70 ALCAPA PATIENTS

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Background: Usually, the electrocardiogram of an infant with anomalous left coronary artery from pulmonary artery (ALCAPA) shows signs of anterolateral myocardial infarction like Q waves in I, aVL, V5 and V6 as well as ST changes and T wave inversions in the same leads, and possibly an abnormal R wave progression in the chest leads.
Material and Method: From 1974 to 2016, 84 patients with ALCAPA were seen at our center. We analyzed all 70 existing preoperative ECGs of these patients.
Results: The heart rate was normal in most patients, while some showed tachycardia. The corrected QT time was longer than 440ms in 24 of 70 patients. We found 3 patients with a left bundle branch block and 1 patient with an incomplete block. Q waves were pathologic just in 57% or 73% of the cases in aVL depending on which threshold was considered as pathologic (Table). Remarkably, we found a delayed R wave progression in 22 patients and no R/S ratio >1 in any lead in 22 patients. ST changes were rarely seen. A T wave inversion was present in 74% of the patients in lead aVL.
Conclusion: Even if there are no or very few typical ECG signs for ALCAPA in infants with heart failure, ALCAPA cannot be ruled out. We did not observe the typical ECG findings in all our patients. Therefore we suggest to use echocardiography or angiography to confirm or disprove any suspicion of the diagnosis ALCAPA.

Table 1.

Q wave	**Q I**	**Q II**	**Q III**	**Q avL**	**Q aVF**	**Q V1**	**Q V2**	**Q V3**	**Q V4**	**Q V5**	**Q V6**
dominant in >0.3 mV & >30 ms	29%	0%	1%	57%	1%	0%	0%	0%	9%	26%	20%
dominant in ≥0.3 mV & ≥30 ms	46%	4%	4%	73%	4%	0%	1%	0%	21%	38%	39%
R/S ratio						**R/S V1**	**R/S V2**	**R/S V3**	**R/S V4**	**R/S V5**	**R/S V6**
first precordial lead with R/S ratio >1						7	2	5	13	13	8
						In 22 patients there was no R/S ratio > 1 in any precordial lead					
ST segment	**ST I**	**ST II**	**ST III**	**ST aVL**	**ST aVF**	**ST V1**	**ST V2**	**ST V3**	**ST V4**	**ST V5**	**ST V6**
Elevation	4%	7%	23%	4%	13%	14%	27%	37%	22%	17%	12%
Depression	13%	3%	4%	20%	4%	8%	19%	12%	13%	15%	10%
normal -0.1 mV ≤J + 60 ≤0.1 mV	83%	90%	73%	75%	82%	79%	54%	51%	64%	68%	78%
T wave inversion	**I**	**II**	**III**	**avL**	**aVF**	**V1**	**V2**	**V3**	**V4**	**V5**	**V6**
present in	41%	9%	9%	74%	9%	35%	13%	11%	16%	28%	38%

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P2609 - INCORPORATING PATIENT REPORTED EXPERIENCE MEASURES AND PATIENT REPORTED OUTCOME MEASURES INTO ROUTINE FOLLOW UP OF CHILDREN AFTER NEONATAL REPAIR OF TRANSPOSITION OF THE GREAT ARTERIES

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Background: With decreasing mortality and morbidity after repair of congenital heart disease, there is increasing emphasis on the use of patient reported experience measures (PREMs) and patient reported outcome measures (PROMs) as key indicators of improvement and quality. Our aim was to integrate PROMs and PREMs into the routine follow-up pathway for infants who had undergone repair of transposition of the great arteries (TGA) and to collect some preliminary data.
Methods: Parents of neonates were asked to complete a PREM about their hospital experience on discharge after their surgery. At their 12 month follow-up outpatient appointment parents completed a PROM about their infant – the infant PedsQL, a generic measure of quality of life assessing aspects of physical and psychosocial health. Comparisons were made with published norms for healthy children.
Results: Over a 45 month period 102 infants were discharged after TGA repair and 93 infants were eligible for 12 month follow-up. Parents of 98 infants completed the PREM at discharge (96% response rate), with 53% reporting that they felt moderately or severely anxious and not feeling adequately prepared for going home. PedsQL scores were available for 64 infants (69% response rate) at 12 months and were not different from those of healthy norms (TGA Total score: 83.31; S.D. 11.93; healthy total score: 82.47; S.D. 9.95).
Conclusions: Incorporating PREMs and PROMs collection into routine follow-up of babies who have undergone TGA repair is feasible. Parents valued the opportunity to provide feedback about their experience and importantly, previously unavailable data were collected about quality of life in the first year after surgery indicating excellent outcomes. Areas for improvement in the hospital experience were also identified. However, collecting such data was resource intensive, with specific challenges identified for data collection in outreach clinics, and alternative methods need to be explored.

P2655 - HEART RATE PRESSOR RESPONSE DURING ISOMETRIC HANDGRIP EXERCISE IN CHILDREN WITH CONGENITAL HEART DIESASE AND HEALTHY CONTROLS

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Background & Hypothesis: The exercise pressor reflex activates an increase in heart rate (HR) via mechanically and metabolically sensitive afferents in contracting skeletal muscle. Children with congenital heart disease (CHD) display exercise intolerance compared to healthy control children, but whether an abnormal exercise pressor reflex contributes to the observed exercise intolerance in children with CHD is not known. We tested the hypothesis that the early HR response to isometric handgrip exercise would be blunted in children with CHD versus healthy control children.
Materials & Methods: Twenty-one children with CHD (11 ± 2 years; females = 9) and 20 healthy controls (11 ± 2 years; females = 11) were studied. CHD diagnoses included simple and complex lesions. Children with a pacemaker or heart transplantation were not included in the study. Children performed isometric handgrip exercise for 2 minutes at 30% of maximal voluntary contraction. The early (first minute) and late (second minute) of the HR response was recorded by surface ECG. Children performed the protocol twice and responses were averaged. A 6-minute walk test was performed to assess exercise capacity. Analyses were completed using unpaired t-tests with $P < 0.05$ being significant. Data are mean ± SD.
Results: 6-minute walk test distance was lower in CHD than controls (523 ± 56 versus 605 ± 79 metres; $P < 0.001$). The early HR response from rest to isometric handgrip exercise at 1 minute was lower in CHD than controls (5 ± 5 versus 10 ± 9 beats/minute; $P = 0.029$). There was no difference in the late HR response from rest to isometric handgrip exercise at 2 minutes between groups (CHD: 10 ± 7 versus controls: 11 ± 8 beats/minute; $P > 0.05$).
Conclusions: A blunted exercise pressor reflex during early exercise may contribute to the pathophysiology of exercise intolerance in children with CHD and low exercise capacity.

P2696 - CONGENITAL HEART BLOCK AND SINGLE INSTITUTIONAL EXPERIENCE WITH EPICARDIAL PACING IN CHILDREN IN A LOW MEDIUM INCOME COUNTRY

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Introduction: Epicardial pacemaker systems are still preferred in many centers than the endocardial pacemaker systems for the young children and infants <10 kilograms because of the increased risk of venous occlusion, associated complex congenital structural cardiac anomalies with limited access, right to left shunts and increased risk of thromboembolism, small size and somatic growth.On the other hand, the epicardial approach is invasive and the epicardial leads are associated with high rate of fractures, increased pacing threshold and reduced generator longevity.
Methods: A retrospective review of cases requiring permanent epicardial pacing presenting to the Division of Paediatric Cardiology at the Chris Hani Baragwanath Academic Hospital between 1992 and 2015. There are no dedicated pacemaker follow up facilities for the children and onsite surgical cardiothoracic services. Data related to demographics, indications, diagnosis, associated structural heart anomalies, pacemaker system, surgical approach, lead and generator longevity, mortality and follow up was collected.
Results: Forty five patients (females,61.1%)underwent insertion of permanent epicardial pacemaker and the majority(55.6%)were diagnosed with congenital complete heart block. Median age at first implant in patients with congenital complete heart block was 3 months (0.099-144 months,IQR = 0.67-69). The median weight was 8.1 kilograms (2.5-36 kilograms,IQR = 3.3-17.2). The other indications were postoperative complete heart block in thirteen cases (28.8%)and sinus node dysfunction in seven cases (15.6%). Associated structural congenital heart anomalies were

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found in twenty patients(44.4%). Fifty nine pacemaker generators were placed either in the pleural cavity or rectus sheath via the antero-lateral thoracotomy or median sternotomy and seventeen pacemaker generators (37.8%) were revised due to battery depletion (mean = 7.16 ± 2.85 years;3.0-14.75 years. Twenty one epicardial leads were replaced (mean = 5.13 ±2.53 years;0.008-9.83 years). Overall mortality was 26.7% and related to the pacemaker in one patient.Mean follow up was 6.89 ±5.09 years (0.5-17.33 years).
Conclusion: Congenital complete heart block is the commonest indication for epicardial pacemaker insertion in children .Lead malfunction and battery depletion were common reasons for revision of the pacemaker system.

P2704 - EVALUATION OF THE RESPONSE TO THE CARDIOPULMONARY TEST OF EFFORT IN UNIVENTRICULAR PATIENTS

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Introduction: It is known that patients with complex cardiac structural alterations have reduced exercise capacity in relation to simple heart diseases. This fact can be explained both by residual hemodynamic defects and by the lack of physical conditioning.
Goals: The objective of this study was to analyze the exercise capacity of children and adolescents who were submitted to Glenn or Fontan procedure and to compare with no heart diseases controls.
Methods: Were included all univentricular patients submitted to cardiopulmonary testing between March 2005 and July 2014. The controls were selected from a sample of children submitted to the same test to assess physical activity during the same period, matching for gender and age. Demographic data and variables of the cardiopulmonary exercise test were compared between the two groups.
Results: There was no statistical difference between the two groups regarding age, height, BMI, heart rate and resting systemic blood pressure, as well as vital capacity. The group of univentricular patients presented a significant reduction of the maximum VO2 (77.9%) in relation to the controls (101.7%) ($p < 0.001$). There was no difference between the respiratory reserve and the maximum oxygen pulse. However, the univentricular group presented lower PET CO2 and higher CO2 equivalent (VE/CO2) at peak exercise ($p = 0.001$), as well as lower initial and final oxygen saturations in relation to control ($p < 0.01$).
Conclusion: Univentricular patients present decreased exercise capacity, which is related to cardiocirculatory limitation. This decreased exercise capacity is not determined by the pulse of O2

P2707 - THE PREDICTIVE VALUE OF PHENOTYPIC FEATURES OF CARDIAC PATIENTS FOR FISH TESTING FOR THE 22Q11.2 DELETION SYNDROME AT RED CROSS WAR MEMORIAL CHILDREN'S HOSPITAL CAPE TOWN SOUTH AFRICA

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Background: Chromosome 22q11.2 deletion syndrome (22qDS), an autosomal dominant disorder, is the most common microdeletion syndrome. More than 180 phenotypic features are associated with 22qDS and this wide phenotypic variability may make it difficult to know when to confirm 22qDS by FISH (fluorescent-in-situ-hybridisation) testing. Its diagnosis is important, since it may have serious implications for the patient's management and prognosis. Óskarsdottir's score is commonly used to suggest the need for FISH testing for 22qDS, but a recent study done at our hospital found its criteria to have a positive predictive value of only 14%! This may be due to the fact that Óskarsdottir's study was based on the phenotypic features of a Caucasian study population, in Sweden, while our population is largely non-Caucasian. We sought to describe which phenotypic features in our patient population would best suggest the need for FISH testing for 22qDS.
Methods: A retrospective review of the clinical case notes for the phenotypic features of patients, who had had FISH tests done after the clinical suspicion of 22qDS, was performed. The clinical triggers to suggest FISH testing of 144 patients (72 FISH-positive 22qDS vs 72 FISH-negative patients) were reviewed.
Results: The four most common phenotypic triggers for testing FISH-positive patients were congenital heart disease (CHD), failure to thrive, "soft" dysmorphic features and developmental delay. Younger children presented frequently with CHD, while older children presented with developmental delay and dysmorphic features. The (combined) positive predictive value (PPV) for these 4 most common triggers was 76.4%, far better than the 14% attained by the Óskarsdottir score in our population.
Conclusion: It is clear that non-Caucasian populations have some unique phenotypic expressions of 22qDS that require different, population-specific diagnostic criteria to suggest performing FISH testing for a confirmation of a diagnosis of 22q11.2 deletion syndrome.

P2743 - CARDIAC SCREENING PROGRAM FOR HIGH SCHOOL ATHLETES IN BERGEN COUNTY A FEASIBILITY AND UTILITY STUDY

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Background: Sudden cardiac death during intense physical activity is a rare but devastating event in children and young adults. Various studies have evaluated the utility and predictive value of cardiac screening programs in high school athletes. We report the results of a cardiac screening program for high school athletes in Bergen County.
Material and Methods: The study population consisted of high school athletes aged 13 to 16 years from local high schools in Bergen County. Study participants filled out a questionnaire based on existing guidelines for sports participation. Salient points included cardiac symptoms, growth and development, past medical history and family history. Informed consent was obtained for exam and screening tests. Limited physical exam included vital signs and calculation of a BMI. Students then underwent a 12-lead electrocardiogram and complete echocardiogram. Study results were interpreted by a pediatric cardiologist, who recommended appropriate follow up. This study analyzes the results of the complete cardiology evaluation on follow up and looks at the predictive value of the screening program.
Results: A total of 450 students between the ages of 13 and 16 participated in the screening program. 104 were flagged as abnormal on the screening based on electrocardiogram or

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echocardiogram, and were advised a complete evaluation by a pediatric cardiologist. Common diagnoses included bicuspid aortic valve, mitral valve prolapse, patent foramen ovale and sinus bradycardia. Our study is ongoing, and will compare the results of the complete cardiology evaluation with the screening results.
Conclusion: Although routine cardiac screening of high school athletes is not recommended, our study aims to look at the feasibility and predictive value of a local cardiac screening program because of the devastating effects of sudden cardiac death on families of young athletes, and potential benefits of prevention.

P2782 - EPIDEMIOLOGICAL AND CLINICAL PROFILE OF CONGENITAL HEART DISEASE IN A REGIONAL MATERNITY CENTER OF TUNISIA

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Background: Congenital heart defects have an estimated worldwide incidence of 6-8 per 1000 live births. Given the frequence of consanguinity in our country, this incidence should be higher in Tunisia.
Objective: The aim of this work was to analyse the epidemiological and clinical characteristics of congenital heart disease among hospitalized newborns in the regional maternity center of Monastir city.
Materials and Methods: This is a retrospective study of patients with cardiac conditions seen at the maternity center over 5 years 8 months realized in our service (from 1 January 2010 to 31 December 2014).
Results: During the study period, 150 cases were analyzed among which 79 summers were selected. The sex ratio of 1.25. Consanguinity was found for 12 couples and congenital heart disease history was found in 4 cases. Prenatal diagnosis was made in 17 cases at an average term of 29 weeks [GA 16-37]. The average age at admission of those with postnatal diagnosis was 16h30 minutes [1 h - 330 h]. After birth, the discovery of congenital heart disease was accidental in 31 cases. In the remaining cases, the most frequent reasons for hospitalization were cyanosis (18 cases) and neonatal respiratory distress (17 cases). Most of children had congenital heart disease involving the right ventricular outflow tract and association with a malformation syndrome was found in 19 newborns with a predominance of down syndrome (6 cases). The Prostine® was indicated for 22 infants and 8 newborns underwent surgery. The overall mortality rate was 38% (30 cases) with an average death age of 13-day [1-75 days].
Conclusion: To improve the management of congenital heart disease we must ameliorate the antenatal diagnosis, which still insufficient in our country, and planning of delivery at risk in a center equipped with appropriate materials and human skills.

P2794 - THE QUOTIENT OF SINUS VALSALVAE AND SINOTUBULAR JUNCTION – AN ADDITIONAL AND INDEPENDENT PARAMETER TO CLASSIFY THE AORTIC AFFECTION OF PEDIATRIC PATIENTS WITH MARFAN SYNDROME

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Objectives: Marfan-syndrome (MFS) is an autosomal dominant connective tissue disorder with a variable clinical presentation. One of the main causes of mortality is transformation of the cardiovascular system. The measurement of sinus valsalvae aortae (SV) in relation to body surface and age (Roman et al) is the common marker to classify the risk for subsequent aortic complications over time. An independent tool for the evaluation of aortic dilation could be the measurement of the quotient of sinutubular junction (SJ) and SV. This additional criteria could be very suitable for a possible therapeutic decision irrespective of age and percentiles. In a first step we compared the SJ/SV quotient of children with and without MFS.
Methods: We investigated 381 pediatric patients (11.4 ± 5.5y) with suspected MFS, 141 patients of this group have diagnosed MFS (11 ± 5.6y). The quotient of SJ and SV was analyzed at first consultation of patients with diagnosed MFS (115patients) and suspected MFS (204patients). Patients with MFS younger than one year and patients without confirmation of MFS and enlargement of the SV (z-score: + 2) were not included in the study.
Results: The mean ± standard deviation was 0.78 ± 0.1 in the group of patients with diagnosed MFS and 0.83 ± 0.09 in the group of patients without MFS. This is a significant difference ($p \leq 0.005$).
Conclusions: There is a significant difference between the quotient of SJ/SV in patients with and without MFS, therefore this new parameter could be an additional tool to facilitate therapeutic decisions. In this first evaluation the new quotient appears to be an age and body surface independent parameter, but more investigations with a larger sample size and over time are needed to analyze whether the parameter is influenced by any other factors, for example whether it changes during adolescence.

P2811 - FOETAL ECHOCARDIOGRAPHY REASONS FOR REFERRAL AND PREVALENCE OF CONGENITAL HEART DISEASE

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Introduction: The prevalence of childhood congenital heart disease is estimated at 8 per 1000 live births. With the advent of foetal echocardiography (FE), early diagnosis of congenital heart diseases became possible, allowing adequate maternal follow-up and planning of birth in reference centres in paediatric cardiology. However, there is still no clear evidence of criteria for foetal echocardiography in the literature, with many exams performed without precise indications. This study aims evaluate relationship between the foetal diagnoses of congenital heart disease and the indications of the examination.
Material and Methods: Retrospective analysis of database of 11702 FE performed in 10921 patients at referral service from January 2004 to August 2016. The first examination of each patient was selected according to the reasons for referral and diagnostic findings.
Results: The main reasons for foetal echocardiography were: check-up (78.0%), a suspected CHD on US (%) maternal risk factor (16.0%), foetal risk factor (6.4%). A normal FE was 96.1%. Among the patients with alterations, arrhythmias (9.7%), complex heart disease (23.9%) and simple heart disease (65.9%) were found. Among the diagnosed heart diseases, the main were: ventricular septal defect (46.4%), coarctation of Aorta (6.9%), atrioventricular septal defects (6.2%). The reasons for referral that presented the

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highest frequency of positive diagnosis for congenital heart disease were: suspected heart disease (60%), fetal risk factor (9%). The variable maternal age didn't change the prevalence of positive diagnoses for congenital heart disease.
Conclusion: The main referral reason for a FE was a check-up. This indication, however, yield only 2,2% of positive diagnosis. Conversely, 60.4% of patients referred due to an abnormal obstetric ultrasound presented with some abnormality And the fetal referrals, particularly due to new indications are leading to detection rates close to 10%. This highlight, the need to reinforce the indications for FE based on more specific criteria.

P2838 - A RARE CAUSE OF HYPERTENSION IN A 7 YEAR OLD BOY

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Background: Arterial hypertension in childhood is more likely to be secondary, but a differential diagnosis may be a challenge.
Case report: A previously healthy 7-year-old boy was referred to a pediatrician due to burning pain in palms and feet lasting for a week. The pediatrician detected high values of blood pressure up to 160/120 mm Hg and sent the boy to the hospital. On admission high blood pressure and tachycardia were found, the skin over the affected area was unchanged, no abnormalities in neurological examination were detected. During the observation in cardiology department he was irritable, suffering of severe pain non-responsive to analgesics, his blood pressure and heart rate were consistently high, temperature normal. Laboratory data did not revealed any abnormalities. Blood pressure and heart rate were constantly elevated, mean values 165/125 mm Hg and sinus rhythm 160 bpm respectively, without a variability during 24-hour monitoring. Shortly after admission he presented first in his life seizures attack without the blood pressure rise and a good response to midazolam. The blood and urine test showed elevated catecholamine metabolites, elevated aldosterone concentration and renin plasma activity. Imaging (ultrasound, CT, MRI) of brain, spine and abdomen showed no pathological masses. Samples of blood, urine and hair were negative for mercury poisoning. The acute autonomic neuropathy was confirmed by negative sympathetic skin response test. Nerve conduction studies were normal. The treatment with intravenous immunoglobulin, calcium blocker, beta-blocker and ACEI was introduced with gradual normalization of blood pressure and heart rate. Gabapentin was used for control of neuropathic pain with good result. All symptoms released during 3 months follow-up.
Conclusions: The described case reiterates the need for careful and open-minded approach to the patients with hypertension. Acute autonomic neuropathy should be considered as a cause of hypertension.

P2841 - REDUCING INTERESTAGE HOME MORTALITY IN A DEVELOPING COUNTRY A HIGH RISK AMBULATORY APPROACH

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Background: During the inter-stage period there is controversy about the possibility of outpatient follow-up, due to high risk of death at home. However, keeping a patient hospitalized for months is also not a good solution. The literature has shown a reduction in home mortality using Home-Care or telemedicine, but this resources are unavailable for many patients from developing country due to cost issues. We developed an option of closely following these patients through a specialized ambulatory service.
Materials and Methods: The goal of our study was to evaluate the impact of a specialized ambulatory follow-up in home mortality of children in inter-stage period. We retrospectively analyzed charts from patients who underwent a palliative procedure in the neonatal period or had a complex cardiopathy that required close follow-up before the first procedure. These patients were discharged from the neonatal admission between jannuary 2015 and December 2016 and followed up in our specialized ambulatory. They were evaluated with close returns between 7 and 15 days, serial echocardiograms, oximetry, adjustment of medications and feeding.
Results: A total of 134 patients were followed in the period. Four patients died at home (0.03%). Currently, 76 patients have already undergone the definitive surgical procedure, 14 of whom remain hospitalized. Twelve patients died in the post-operative period and 50 patients no longer require closely follow-up and were switched to a conventional ambulatory. In addition, 54 patients are still being followed up in our ambulatory.
Conclusions: Inter-stage is a critical period in the management of this patients. Currently in Brazil we do not have enough resources to follow these children with Home-Care or telemedicine, so the proposal of specialized ambulatory follow-up with close returns and serial echocardiograms presents as an alternative to lower the mortality rate in this period.

P2842 - FUNCTIONAL CAPACITY IN POSTOPERATED TETRALOGY OF FALLOT PATIENTS ERGOSPIROMETRIC RESULTS ACCORDING TO THEIR PATHOPHYSIOLOGY

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Background: Postoperated Tetralogy of Fallot patients (PTF) show decreased exercise capacity (ExC) (represented by VO2) and increased VE/VO2 slope. Both parameters are factors of morbidity and predictors of death and hospitalitation. Right ventricular dysfunction and residual pulmonary stenosis or regurgitation severity play a significant role in the evolution of these patients.
Objectives: To correlate the pathophysiology and anatomy in pediatric PTF population with its ergospirometric parameters.
Methods: Retrospective observational study of 25 PTF (mean age 12 ± 3.2 years, weight 43.2 ± 14.55 kg, height 146.7 ± 14.4 cm, 58% male). Inclusion criteria admitted pulmonary atresia with VSD. Ramp treadmill ergometry (Bruce protocol) was performed with expired gas in all cases.
Results: Submaximal exercise tests limited by symptoms were performed, clinically and electrically negative in all cases, with no significant arrhythmias. 56% showed a normal spirometric pattern and 20% restrictive. 32% reached 85% predicted HR ($79,6 \pm 9,3$%. Rest HR 89 ± 14 bpm, maximal HR 170 ± 17 bpm (HRR 81 ± 22 bpm). SBP/DBP at rest was $97 \pm 10/57 \pm 8$ mmHg,

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maximum of 125 ± 10/71 ± 8 mmHg. DP was 20938 ± 2817. CPET showed decreased maximal VO2 (79,5 ± 10,2% predicted), with normal O2 pulse(101 ± 18,5% predicted). The rise in Vslope and CO2Eq at VAT (33,4 ± 8,3 and 34,9 ± 6,8 respectively) botht at the high limit of normality could translated ventilatory inefficiency. Respiratory reserve 42 ± 14,6 (normal).
Conclusions: CPET provides prognostic information in these patients, and can determine risk factors. Certain values of VO2 and V slope are associated with specific pathophysiological situations: greater V slope is found in pulmonary stenosis, lower VO2 when right ventricular dysfunction and greater pulmonary regurgitation occur. These data are relevant in the complex ergospirometric interpretation in congenital heart disease, within the limitations of a small sample size.

P2844 - ANDERSEN TAWIL SYNDROME A TOO LATE DIAGNOSIS

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Background: Andersen-Tawil syndrome (ATS) is an autosomal-dominant disorder characterized by ventricular arrhythmias, periodic paralysis, distinctive facial-skeletal anomalies and developmental abnormalities. The affected genes is KCNJ2. Cardiac manifestations include Premature Ventricular Contraction (PVC),Q-U prolongation, prominent U waves,ventricular tachycardia. Family history of sudden cardiac death sometimes is not known.
Case: We present a 42 year old woman with history of Psychological negligence abuse in childhood and eating disorder. Father died 2 years ago. Mother is alive. First of 3 sisters. No past medical history. Problems during childhood, poor social relationships. During first hospitalization episode in Psychiatric ward it was documented a BMI under 15, Mixed personality disorder with autolytic threats; a consultation to clinical genetics unit was done, due of dysmorphic phenotype. Physical exam with small stature, small lower jaw, low-set ears, widely spaced eyes, clinodactyly were demonstrated. Cardiovascular exam,no murmurs were found. An ECG with a slightly prominent U wave in V3-V4 leads with a QUc of 660 mseg was found. 24 Hours Holter documented frequent PVC without any VT episode. An Echocardiogram was perfomed without any abnormal founds. The Comparative Genomic Hibridization (CGH) array demonstrated a Delection 17q24.3 in KCNJ2 gen. No antiarrhytmic medication was prescribed. Quetiapine, Paroxetine and Diazepam were prescribed.
Conclussion: ATS is a rare channelopathy, this type of patient present with a wide clinical spectrum from distinctive facial-skeletal anomalies, ventricular arrhythmias, neurocognitive anomalies and periodic paralysis, our patient fullfil most of clinical criteria confirmed by a genetic test. ATS patient has a very high arrhythmia burden, but most frequently asymptomatic, the optimal pharmacological therapy has to be identified.Despite the late timeframe to diagnosis, the management and treatment of this type of patient require a high degree of clinical suspicion and remember to us the importance of a multidisciplinary approach.

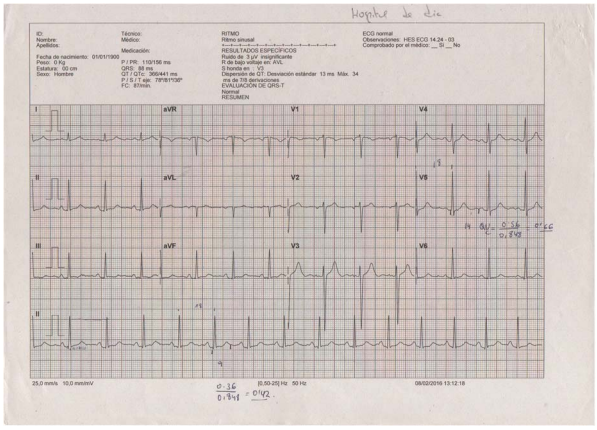

Figure 1.

P2976 - CHALLENGES OF SURVEILLANCE IN CLINIC FOLLOWING COARCTATION OF THE AORTA REPAIR

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Objectives: Identify the monitoring challenges in outpatient department setting post repair of Coarctation of the Aorta.
Methods: 1 year retrospective review of all children seen in clinic with status post Coarctation of the aorta repair. 136 paediatric patients were seen in our clinics at The Evelina Children Hospital from 15th January 2016 to 15th January 2017.We enrolled patients with isolated Coarctation of the aorta, associated atrial or ventricular septal defect and any aortic valve malformation.
Results: For all the 136 patients we recorded weight, height, casual systolic and diastolic blood pressure, descending aortic arch gradient post operative and in clinic, interventricular septum and posterior wall dimensions, anatomy of the aortic valve, timing of diagnosis and age at surgery. For a random subset of this group (n = 69 patients) reliable SBP was available in 62 patients of whom 36.6% had SBP > 95th centile and 16% of them are already under treatment for systemic arterial hypertension. The median age at operation was 13 days and an antenatal suspicion of coarctation was raised in 35%. The systolic blood pressure was significantly associated with the Doppler velocity in the descending aortic arch (r = 0.319, p = 0.012)There was poor correlation between LVPW z scores and SBP z scores (r = -0.104, p = 0.476). Age at operation and hypertension has also positive correlation with statistically significance (r = 0.301 , p = 0.016).
Conclusion: In our subset of repaired coarctation we noticed strong association between SBP and Doppler arch velocity. The prevalence of hypertension following coarctation repair is worryingly high and further evaluation with a control group is warranted.

P3034 - A NEWBORN WITH NOONAN SYNDROME PRESENTED AS RESPIRATORY DISTRESS BY H CMP AND CYSTIC HYGROMA

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Noonan syndrome (NS) is Autosomal dominant (1/1,000-2,500 live birth) characterized by the hypertelorism, ptosis, short neck, low-set-ears, short stature, CHD, and multiple skeletal and

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hematologic et al. Most patient (94%) with positive genotype had known CHD. PTPN11 mutation was noted in 79% of pulmonic stenosis and RAF1 & specific PTPN11 mutation in 12% h-CMP. Children with NS diagnosed with h-CMP before 6month(51%), but rare neonatal presentation.
Case: GA 38wks, 3560 g, NSVD was presented with Cyanosis & tachypnea, which was Not improved with Oxygen therapy by hood tent. Prenatal USG showed the short femur Vital sign : HR 148 /min, RR 68/min, BT :36.5°C, BP : Arm 57/31 mmHg Leg 61/35 mmHg WT 3500gm(50-90pecentile) Ht 47.3 cm(10-50percentile) HC 39.5 cm (>97 percentile) A characteristic configurations of patient were cystic hygroma in neck, flat nose bridge, low set ear and hairline. Mild chest retraction, inspiratory stridor, midsystolic ejection murmur along left sternal border were noted. Cardiac hypertrophy of left ventricle and diastolic dysfunction without left ventricular outflow obstruction was noted on echocardiograhphy. Mechanical Ventilation for severe respiratory distress and then replaced to N-CPAP, continued until 7th day due to CO2 retentions and tachypnea. Oxygen was stopped on the 18th day. Upper airway was not obstructive, and spontaneous resolution of cystic hygroma without surgery. Gene study : negative for PTPN11 gene c.770c > T (p.Ser257Leu) of RAF1 gene. In Neonatal Noonan syndrome, respiratory distress may be appear due to cystic hygroma and h-CMP. Hypertrophic CMP in NS is over represented in RAF1 rather than PTPN11 association, common mutation in patient with PS.The infant require regular detailed follow up for heart and respiratory complication due to hypertrophic CMP and cystic hygroma

ANESTHESIA

P1092 - INCIDENCE OF COMPLICATIONS AND ANESTHETIC MANAGEMENT OF PATIENTS UNDERGOING PULMONARY ARTERY REHABILITATION IN THE CATHETERIZATION SUITE A RETROSPECTIVE REVIEW

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Introduction: Patients with complex pulmonary artery (PA) stenosis often undergo multiple procedures involving balloon angioplasty, cutting balloon angioplasty and stent implantation collectively referred to as pulmonary rehabilitation. We sought to evaluate the anesthetic management and outcomes of PA rehabilitation cases, focusing on known complications.
Methods: Single center retrospective review of patients who underwent PA angioplasty with or without stent placement between October 1, 2010 and October 1, 2015. Data collected included diagnosis, procedure, anesthetic techniques, use of bronchial blockers, pulmonary bleeding, vascular injury detected on chest X- ray, and postoperative course. Additionally, we collected intraoperative hemodynamic data, including right ventricle/femoral artery (RV/FA) pressure ratios.
Results: Data were collected for 108 patients. All patients with single ventricle physiology were excluded. Table 1 compares perioperative data between tetralogy of Fallot (TOF) with pulmonary atresia and other patients. Table 2 illustrates perioperative outcomes and the rate of use of bronchial blockers.
Discussion: Patients with TOF with pulmonary atresia were more likely to experience complications related to PA angioplasty with or without stent placement. Higher RV/FA pressure ratio increased the risk for complications. While use of bronchial blockers in this cohort was infrequent, this may represent the evolution of our anesthetic management of these patients over the study period. Our current protocol is to place a bronchial blocker, either through the lumen of the endotracheal tube or outside, at the beginning of the procedure in patients deemed to be at high risk for pulmonary hemorrhage. The bronchial blocker can be advanced into either lung under fluoroscopic guidance in the event of significant pulmonary hemorrhage or reperfusion injury that interferes.

Table 1. Perioperative data

	TOF + Pulmonary Atresia	Others	P
Number	36	72	
Age (Years)	4.4 ± 4.1	8.2 ± 8.6	0.002
Weight (Kg)	17.1 ± 10.1	29.6 ± 29.1	0.001
Pre procedure RV/FA ratio	1 ± 0.1	0.8 ± 0.3	0.0002
Number of branches dilated	4 ± 1	3 ± 2	0.00015
Post procedure RV/FA ratio	0.9 ± 0.1	0.6 ± 0.2	<0.001
Case time (Minutes)	278.4 ± 84	230.4 ± 82	0.008

Table 2. Perioperative outcomes

	Mild bleeding in endotracheal tube	Severe bleeding in endotracheal tube	Vascular injury detected by X ray
Number	10	8	10
Anesthesia placed bronchial blocker prior to interventions (Number)	4	1	1
Remained intubated (Number)	2	7	0
CTICU admission (Number)	5	8	2

P1103 - EARLY EXTUBATION EXPERIENCE AT A REGIONAL HEART CENTER IN RUSSIA

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Introduction: Medical mission trips are a very rich environment to exchange experiences between the visiting and the home teams.

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Assessing the success of a change in practice based on such experience can be a very practical tool in establishing the real impact of such experience. The concept of early tracheal extubation after repair of congenital heart disease utilizing cardiopulmonary bypass (CPB) has been adopted as a safe technique in many heart centers across North America. In this retrospective review, we evaluated the progress of early tracheal extubation in children undergoing surgery for congenital heart disease utilizing CPB in a Heart Center program in Russia after a multidisciplinary team visit from the United States.
Methods: A retrospective review of extubation data for children undergoing congenital cardiac surgery at a major heart center in eastern Russia between August 1st 2016 and September 30th 2016. As a control group, we collected data between January and December 2015. Data included age, weight, gender, type of lesion, CPB time, aortic cross clamp time, complications, cardiac intensive care unit (CTICU) length of stay (LOS) and hospital LOS.
Results: A total of 312 were included in the study cohort. We excluded 53 patients who had thoracic surgeries not requiring CPB. A total of 259 patients were included in the final cohort (221 prior and 38 after implementing the early extubation protocol). Table 1 illustrates the demographics and perioperative outcomes
Discussion: Our early results demonstrate that early extubation can be achieved safely at a rate (86.8%) that is very comparable to many centers in North America. This success has resulted in reduction of both the CTICU and hospital LOS. Validation of such results is necessary over a long term to illustrate the ability to reproduce such results with its potential clinical and economic impact.

Table 1. Demographic and perioperative differences.

	Early Extubation Group	Control (Prior to early extubation protocol)	P
Total number of CPB patients	38	221	
OR extubation (number + %)	33/38 (86.8%)	0/221 (0%)	<0.00001
Age (months)	32 ± 45	42 ± 91	0.28
Weight (Kg)	11.3 ± 10	13 ± 15	0.34
Gender (male/female)	19 M/19 F	110 M/111 F	0.88
CPB time (minutes)	59.6 ± 29.6	70 ± 41	0.0537
Aortic cross clamp time (minutes)	41 ± 20.6	42 ± 25	0.768
Incidence of complications (number + %)	5 (13.2%)	57 (25.8%)	0.049
ICU LOS (days)	2 ± 2	5 ± 3	0.000003
Hospital LOS (days)	12 ± 3	15 ± 9	0.00001

P1693 - ANESTHESIOLOGIC MANAGEMENT FOR PERCUTANEOUS POTTS CREATION IN PEDIATRIC PATIENTS AFFECTED WITH SUPRASYSTEMIC PULMONARY HYPERTENSION

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Patients with suprasystemic PAH and NYHA class III –IV have poor prognosis, limited therapeutic options and are extremely fragile. We describe our modified anesthetic management for percutaneous Potts shunt creation in a pediatric population. Anesthesia has been modified to ensure safe induction, close monitoring of the patients during this challenging procedure (Creation of anastomosis between descending aorta and left pulmonary artery with covered stent after radiofrequency-guided vessel perforation) and weaning from anesthesia without rebound PH. We report our management for 8 patients. In case of venous access, arterial and second venous access were obtained after ketamine injection and local lignocaine administration, when no access was usable, sevoflurane was used to place peripheral IV and arterial lines. One access was used for epinephrine and norepinephrine administration. Once lines were secured, deep anesthesia was carried out with alfentanil and etomidate. In case of BIS index >50, propofol (Extremely titrated) was given if mean arterial pressure was >65 mmHg, ketamine was used otherwise. General anesthesia was maintained with sevoflurane, alfentanil and atracurium and patients were ventilated with 100% inspired fraction of oxygen and 20 ppm of nitric oxide. Perforation and stent placement were generally uneventful. 2 patients presented extremely low cardiac output after dilatation of the stent and effective Potts shunt creation due to acute off-loading of the left ventricle. VA ECMO was placed during resuscitation. Patients died because of brain damage. One patient has preventive VA ECMO after uneventful induction because of severe biventricular function; he died 1 month after the procedure because of ARDS. The remaining 5 patients are alive. Mean intubation duration and mean inotropic support were similar: 1.2 days; mean ICU stay was 2.4 days. Ventricular function and functional status improved in all patients. Our modified anesthetic management proved to be helpful in this challenging procedure in the vulnerable population.

Table 1. Patient's data and outcome.

p	W (Kgs)	H (cm)	A (Year)	NYH A class	ECMO (Y/N)	Early death < 7d(Y/N)
1	34	145	13,3	3	N	N
2	78	157	17	3	N	N
3	33	132	9,4	3	Y	Y
4	36,8	131	6,7	3	N	N
5	17,5	108	5,8	3	Y	N
6	26,8	133	10,7	3	N	N
7	45	168	14,3	4	Y	N
8	35	136	8,9	3	N	N
%						12.5
Mean	36,8	138,75	10,7			
Range	17,5-78	108-168	5,8-17			

p	Death <30d (Y/N)	Death Before HD (Y/N)	Int.d	Inot r d	ICU d.	NYHA class after Potts
1	N	N	1	2	3	2
2	N	N	1	1	3	3
3	Y	Y	2	2	2	Un.
4	N	N	1	0	1	2
5	Y	Y	8	8	8	Un.
6	N	N	1	1	2	1
7	N	Y	40	40	40	Un.
8	N	N	2	2	3	3
%	25	37.5				
Mean			7	7	7,75	
Range			1-40	0-40	1-40	

Key to symbols. P = patient; W = weight; H = height; A = age; ECMO = Extra Corporeal Membrane Oxygenation; NYHA = New York Heart Association; Y = yes; N = not; d = day; Int. = Intubation; Inotrop. = Inotropes; ICU = Intensive Care Unit; Un. = Unknown.

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P1754 - IMMEDIATE AND SHORT TERM OUTCOMES OF TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION REPAIR AT A QUATERNARY CARE CENTRE IN INDIA

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Background: Strategic perioperative management of TAPVC in recent years has markedly improved the outcomes in developing countries like India. We share our retrospective short term outcome of TAPVC.
Methods: All TAPVC patients (n = 166) operated between June 2012 and September 2016 at our centre were included. Postoperative outcomes evaluated as 30 day mortality and morbidity were analysed amongst the various TAPVC subsets.
Results: Of 166 TAPVC patients, TAPVC was classified as Supracardiac in 91 patients (54.8%), cardiac in 18 patients (10.8%), Infracardiac in 45 patients (27.1%), and mixed in 12 patients (7.2%). Obstructed variants (n = 88) and unobstructed ones were (n = 78). 106:60 M:F, the mean age (month) was 2.75 ± 7.14 mean BSA calculated was 0.24 ± 0.07. No direct correlation of duration of CPB (148.87 ± 119.68) and ACC (68.65 ± 25.97) on any of the morbidity parameters. Duration of ventilation is significantly higher in Infracardiac (97.29 ± 82.67) and Mixed type (56.08 ± 38.39) TAPVC in comparison with Supracardiac (56.63 ± 73.18) ($p < 0.004$) and Cardiac type (42.93 ± 31.31) ($p < 0.009$) TAPVC. The mean duration of ionotrope in obstructed group (5.5 ± 2.15) was significantly higher as compare to unobstructed group (3.42 ± 1.84) ($p < 0.001$). We have also found statistically significant difference in the mean duration of ventilation (hrs) (85.17 ± 80.94 (66.92-103.42) in obstructed Vs 49.23 ± 60.70 (36.67-62.09) in unobstructed group) and duration of ICU stay (in obstructed (9.64 ± 5.96) with CI of (8.30-10.98) and 6.29 ± 5.12 (5.21-7.38) in unobstructed group p(<0.001)). There was a negative correlation between BSA of patient and duration of inotrope, ventilation time, length of ICU stay ($r = -0.313$, 2 tailed $p < 0.001$). Total mortality in our series was 9/166 which was 5.4% Risk factors for mortality in our study included lesser BSA (0.24 ± 0.07), type and Obstructed variant of TAPVC.
Conclusion: Prolonged use of ionotropes, longer ventilation time and ICU

P1755 - ONE AND HALF VENTRICLE REPAIR IN CASE OF ASD VSD AND HYPOPLASTIC RIGHT VENTRICLE PERIOPERATIVE PROBLEMS AND MANAGEMENT

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'One and a half ventricular repair' is a surgical option for congenital cardiac anomalies characterized by a hypoplastic but potentially or partially usable right ventricle which reduce early risk and late failure while still achieving separate pulmonary and systemic circulations. We describe successful outcome Managing a 7month old female with failure to thrive diagnosed of Large VSD and ASD with hypoplastic Right ventricle and small tricuspid z score -2.37. She was candidated for vsd closure but intraoperatively difficult weaning due to significant right to left shunting across ASD so plan was made to close ASD asd In an effort to reduce the volume load of the ventricle glenn shunt was also done. Postoperative management was challenging maintaining PA pressure surges with ventillatory strategies and pulmonary vasodilators. She had a complicated postoperative course of low cardiac output and maintaining hemodynamics was important key factor. Thus so called 'one and half ventricle repair' is to achieve a physiological correction by separating the pulmonary circulation from the systemic one, 'unloading' the insufficient RV at thetime of the correction,but most challenging is postoperative management of these subsets. We emphasize early anticipation and strategical postoperative management of such complicated repairs

P1882 - FEASIBILITY OF AUTOLOGOUS INTRAOPERATIVE BLOOD COLLECTION AND RETRANSFUSION IN SMALL CHILDREN WITH CONGENITAL HEART DEFECTS UNDERGOING CARDIOPULMONARY BYPASS

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Introduction: Transfusion of allogeneic blood products is common in infants undergoing cardiac surgery and cardiopulmonary bypass (CPB) although it is associated with an increased risk for adverse events. Autologous intraoperative blood collection and re-transfusion (AIBC) is a proven method of blood preservation, but has not been extensively practiced in smaller children. In this study we compare two groups of children weighing <10 kgs, one that received AIBC and one that did not, in terms of the efficacy and safety of this practice.
Methods: After IRB approval, we retrospectively reviewed the medical records of children weighing <10 kgs who underwent CPB between 1/1/2014 and 1/31/2016. Control patients were chosen in a 1:1 matched design based on three criteria: preoperative hematocrit (HCT), procedure, and patient body weight. For patients in both groups, we collected corresponding and pertinent demographic and surgical data, clinical outcomes and certain adverse events.
Results: Patient demographics (Table 1) and preoperative laboratory values (Table 2) were similar between the two groups. Study patients tended to require more inotropic support prior to CPB than control patients. Study patients also had a greater prolongation of immediate postoperative coagulation values when compared to the control patients. Despite this, outcome variables were either the same or improved in the study group (Table 3). Specifically, patients in the study group had significantly fewer donor exposures and a shorter ICU length of stay.

Table 1. Patient Demographics

	Study (n = 15)	Control (n = 15)	p-value
Age (days)†	238 ± 137	192 ± 124	0.07
Weight (kg)	7.1 ± 1.6	6.7 ± 1.5	0.43
Gender male (% male)	8 (53)	9 (60)	0.99
Procedure category (%)			
1	1 (7)	1 (7)	
2	10 (67)	12 (80)	
3	4 (27)	2 (13)	
Redo sternotomy (%)	7 (47)	12 (80)	0.13
Single ventricle (%)	11 (73)	12 (80)	0.99
Cyanosis (%)	15 (100)	15 (100)	
Preop transfusion (%)	0 (0)	0 (0)	
Pre-CPB inotropic support (%)	7 (47)	3 (20)	0.25
CPB time (min)	90 ± 56	120 ± 87	0.28
Aortic cross-clamp time (min)	32 ± 39	41 ± 54	0.57
Lowest temperature (°C)	33 ± 5	31 ± 6	0.17
MUF (%)	8 (53)	13 (87)	0.13

⋆ : Mean ± SD or N (%) are reported.
†: Non-parametric paired Wilcoxon Rank Sum Test utilized.

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Discussion: AIBC is a viable option for children weighing <10 kgs. While the immediate postoperative coagulation values were more prolonged in the study group, they still fell within an acceptable range and did not result in greater chest tube output or extra transfusions. Importantly, fewer donor exposures could significantly benefit patients who need future surgery or cardiac transplantation. Our data support the concept that AIBC is an efficacious and safe practice in patients <10 kgs.

Table 2. Preoperative and Postoperative Laboratory Values.

	Study (n = 15)	Control (n = 15)	p-value
Preoperative			
Hgb (g/dL)	17.2 ± 1.8	17.4 ± 1.5	0.58
Hct (%)	51.5 ± 5.6	53.4 ± 4.2	0.10
Platelet (x 10^3/μL)	348 ± 140	293 ± 119	0.12
PT (sec)	14.3 ± 1.4	14.6 ± 1.1	0.24
INR	1.1 ± 0.1	1.1 ± 0.1	0.82
PTT (sec)	31.7 ± 4.7	33.5 ± 3.2	0.13
Fibrinogen (mg/dL)	245 ± 54	267 ± 59	0.42
Postoperative			
Hgb (g/dL)	14.3 ± 1.9	13.1 ± 1.9	0.09
Hct (%)	41.8 ± 5.3	38.8 ± 5.4	0.11
Platelet (x 10^3/μL)	186 ± 72	259 ± 78	0.01
PT (sec)	18.7 ± 2.6	17.1 ± 1.3	0.04
INR	1.6 ± 0.3	1.4 ± 0.1	0.03
PTT (sec) †	49.0 ± 42	36.9 ± 9.2	0.07
Fibrinogen (mg/dL)	210 ± 101	364 ± 90	< 0.001

⋆: Mean ± SD are reported.
†: Non-parametric paired Wilcoxon Rank Sum Test utilized.

Table 3. Patient Outcomes

	Study (n = 15)	Control (n = 15)	p-value
24 hour CTO (ml/kg)	26 ± 11	21 ± 14	0.33
Total volume transfused (mls/kg)	50 ± 50	68 ± 43	0.24
Total donor exposures (n)	3 ± 3	6 ± 2	< 0.001
Duration mechanical ventilation (hours)†	29 ± 48	50 ± 133	0.25
Length ICU stay (days)†	3 ± 3	7 ± 11	0.02
ECMO, N (%)	0 (0)	0 (0)	
In-hospital mortality, N (%)	0 (0)	0 (0)	

⋆ : Mean ± SD or N (%) are reported.
†: Non-parametric paired Wilcoxon Rank Sum Test utilized.

P1894 - RED BLOOD CELL TRANSFUSION AS A PREDICTOR OF OUTCOME AFTER CARDIAC SURGERY IN INFANTS LESS THAN SIX MONTHS OF AGE

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Introduction: Red blood cell transfusion (RBCT) during pediatric cardiac surgery benefits patients when anemia becomes sufficiently severe to decrease oxygen delivery and when acute blood loss causes hemodynamic instability. Yet, RBCTs are not without risk and, in general, are associated with poorer postoperative clinical outcomes. In this study, we examine the incidence of RBCT and its association with postoperative clinical outcomes in infants ≤6 months of age undergoing cardiopulmonary bypass (CPB).

Methods: We retrospectively reviewed the medical records of 666 infants ≤6 months of age who underwent CPB at our institution over a 3 year period. Demographic and surgical data, intraoperative and postoperative RBCT volumes, and several postoperative clinical outcomes were collected. We analyzed the data for associations between RBCT volume and postoperative clinical outcomes after adjusting for disease severity.
Results: Of the 666 total infants, only 3 did not receive an intraoperative transfusion; 455 infants did not receive a postoperative transfusion. The median amount of RBCT intraoperatively was 74 (IQR: 51-125) mls/kg and postoperatively was 28 (IQR: 7-1256) mls/kg. Tables 1 and 2 show our results according to postoperative transfusion status. In our multivariate analysis, significant variables for intensive care unit (ICU) length of stay (LOS) were age ($p < 0.001$), CPB time ($p < 0.001$), intraoperative transfusion volume ($p = 0.01$) and postoperative transfusion as a yes/no variable ($p < 0.001$). For in-hospital mortality, significant variables were CPB time ($p < 0.001$), intraoperative transfusion volume ($p < 0.001$) and postoperative transfusion yes/no ($p = 0.005$).
Discussion: In infants ≤6 months of age undergoing CPB, RBCT is associated with poorer postoperative clinical outcomes. After adjusting for disease severity, the amount of intraoperative transfusion volume and the occurrence of a postoperative transfusion were associated with a longer ICU LOS and in-hospital mortality. Further study is needed to determine if a threshold exists below which the effects of RBCT are not as harmful.

Table 1. Clinical Outcomes.

Outcome	Overall (n = 666)	No post-op transfusion (n = 455)	Post-op transfusion (n = 211)	p-value
Duration of mechanical ventilation (hours)	119 ± 227	83 ± 159	215 ± 329	<0.001
ICU length of stay (hours)	251 ± 495	170 ± 336	438 ± 700	<0.001
Hospital length of stay (days)	21 ± 33	17 ± 28	32 ± 41	<0.001
ECMO	40 (6)	12 (3)	31 (15)	<0.001
Thrombosis (%)	55 (8)	18 (4)	37 (18)	<0.001
Renal failure (%)	12 (2)	0 (0)	12 (6)	<0.001
Infection (%)	65 (10)	33 (7)	34 (16)	<0.001
In-hospital mortality (%)	34 (5)	9 (2)	27 (13)	<0.001

ICU = intensive care unit; ECMO = extracorporeal membrane oxygenation.

Table 2. Patient Demographics.

Variable	Overall (n = 666)	No post-op transfusion (n = 455)	Post-op transfusion (n = 211)	p-value
Age (days)	77 ± 65	83 ± 64	64 ± 66	<0.001
Weight (kg)	4.4 ± 1.4	4.6 ± 1.5	4.1 ± 1.4	<0.001
STAT score (%)				<0.001
1	132 (20)	116 (26)	16 (8)	
2	154 (23)	101 (22)	53 (25)	
3	127 (19)	99 (22)	28 (13)	
4	167 (25)	105 (23)	62 (29)	
5	83 (12.5)	34 (7)	49 (23)	
unassigned	3 (0.5)	0 (0)	3 (2)	
CPB time (min)	114 ± 60	103 ± 49	139 ± 73	<0.001
Ischemic time (min)	58 ± 39	56 ± 35	61 ± 46	0.18
Regional perfusion time (min)	8 ± 19	6 ± 16	14 ± 24	<0.001

STAT = Society of Thoracic Surgeons - European Association of Cardio-Thoracic Surgery Congenital Heart Surgery Mortality; CPB = cardiopulmonary bypass.

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P2001-10 DAY OLD WITH HYPOPLASTIC LEFT VENTRICLE SCHEDULED FOR A NORWOOD PROCEDURE AND BY THE WAY HAS EPIDERMOLYSIS BULLOSA (EB)

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Introduction: Epidermolysis Bullosa (EB) is a rare disease characterized by extremely fragile skin and blister formations in response to friction, trauma, or pressure.1,2 Perioperative management in a cardiac patient with EB requires meticulous care to skin, adhesive dressings, monitors, intubation, and invasive procedures.1,3
Case Presentation: A 10 day old, 3 kg female, with history of heterotaxy, left atrial isomerism, right dominant unbalanced AV canal, hypoplastic left ventricle presented for the first-stage Norwood procedure. EB was diagnosed postnatally based on her father's history of EB and her physical presentation. After multidisciplinary discussions regarding the risks of proceeding with a multi-stage cardiac repair in the setting of EB, the surgery was scheduled. Dermatology was consulted to determine appropriate adhesives for securing critical monitors, invasive lines, endotracheal tube, and the risk of the transesophageal (TEE) probe placement. The patient was intubated preoperatively and duoderm® was placed on her face to protect her skin from the adhesive tape used to secure the endotracheal tube. A preoperative femoral arterial line and a left upper extremity PICC both were covered with a Sorbaview® occlusive dressing. Two additional PIVs were placed intraoperatively, one covered with a Tegaderm™, while the femoral PIV was sutured and covered by a Sorbaview®. An adhesive NIRS (Near-Infrared Spectroscopy) cerebral oximetry probe was placed preoperatively and electrocardiogram leads were trimmed so only adhesive gel remained for contact. One pulse oximetry probe was placed directly on her hand preoperatively and one was placed with a Tegaderm™ attached to the adhesive side so no adhesive material touched her. Lubricant eye ointment was applied followed by a petrolatum gauze cover and subsequent TEE probe placement. Upon completion of the surgery while dressing her open surgical site, removal of an Ioban™ film inadvertently exfoliated the patient's epidermis from her chest and abdomen. Mepilex® Lite and Mepilex® Transfer were applied based on wound care recommendation.
Discussion: There are more than 20 EB subtypes2 and the mouth, pharynx, upper airway, and esophagus may all be involved, resulting in esophageal strictures or increasingly difficult intubations. 3 Unfortunately, our patient has complex congenital heart disease that will require additional surgeries and diagnostic procedures that necessitate invasive monitoring. Therefore, a multidisciplinary approach is essential to reduce the risk of morbidity and mortality from EB or other disease states.1
Conclusion: Adequate perioperative evaluation and preparation are essential in managing congenital cardiac patients with EB2 who will require numerous surgical and catheter-based interventions.
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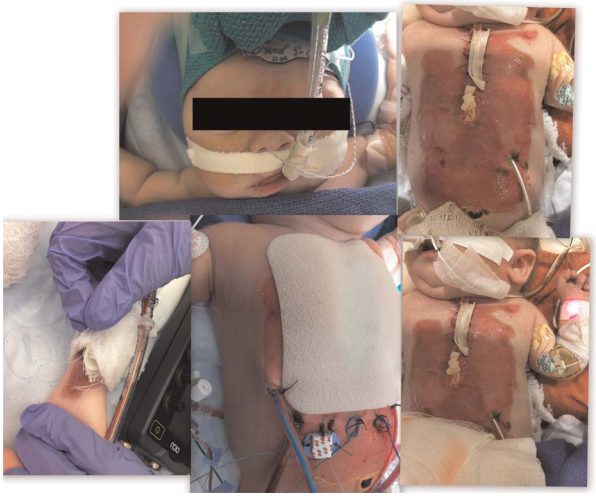

Figure.

P2028 - INCIDENTAL DIAGNOSIS OF CONGENITAL TRACHEAL STENOSIS IN PATIENTS WITH CONGENITAL HEART DISEASE

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Congenital tracheal stenosis (CTS) is a rare anomaly that is difficult to diagnose. The anesthesiologist can be the first to diagnose CTS usually due to an unexpected difficult airway management either upon intubation or in the immediate post-operative period. We present a series of patients with previously unrecognized complete tracheal rings diagnosed in the perioperative and immediate postoperative period. See Table and Figures.
Discussion: Congenital tracheal stenosis is a rare anomaly (incidence of 1:64500)$_1$. It is a reduction of the tracheal diameter by more than 50% due to the formation of complete or near complete tracheal rings$_1$. It can be a neonatal life threatening respiratory anomaly. It is usually associated with congenital heart disease >50% which can make diagnosing these patients very challenging. Congenital tracheal rings are frequently associated with cardiac anomalies including ASD, VSD, CAVC, PDA, vascular anomalies (pulmonary sling) and non-cardiac anomalies like tracheaesophageal fistula, esophageal atresia, Trisomy 21, VATER/VACTERL syndrome, Pfeiffer's syndrome.$_2$ Patient's symptoms range from being asymptomatic to having biphasic stridor with respiratory compromise. Patients with CTS and congenital heart disease can develop pulmonary edema and respiratory distress that can lead to underdiagnosing CTS. Some of these patients present

Table 1.

	Case 1	Case 2	Case 3	Case 4
Age/weight/ Sex	6mo/5kg/F	14mo/9kg/M	5mo/6kg/M	ld/3kg/F
CHD	VSD	TOF	CAVC-Trisomy21	Truncus arteriosus
ETT size	3.5 cuffed	4.0 cuffed	3.5 cuffed	3.0 cuffed
Depth	13.5 cm nares	14cm nares	13 cm nares	9 cm oral
Previous intubation	None	None	None	None
Symptoms post procedure	Desaturation 1 hour post operatively, high ETT on CXR	Failed extubation POD5, stridor	High ETT on CXR, desaturation after attempting to advance ETT	Multiple Failed extubation, stridor
Diagnosis	Long segment CTR	Short segment CTR,LPA sling	Long segment CTR	Mid tracheal CTR, LPA sling
Timing of diagnosis	1 hour post-op	Intraoperatively	1 hour post-op	8 days post-op
Timing of surgical repair after diagnosis	Awaiting repair	Immediately after diagnosis	1 month	Awaiting repair

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early in their neonatal life to the OR for cardiac surgery prior to having developed respiratory symptoms caused by $CTS_{1, 2}$. In the Immediate postoperative period, difficulty ventilating the patient after recovery from the neuromuscular blockage or with attempting to advance the ETT should prompt the anesthesiologist to suspect tracheal narrowing. Diagnosis is confirmed with a bronchoscopic evaluation, cardiac CT and or cardiac MRI may be necessary to evaluate for vascular anomalies.

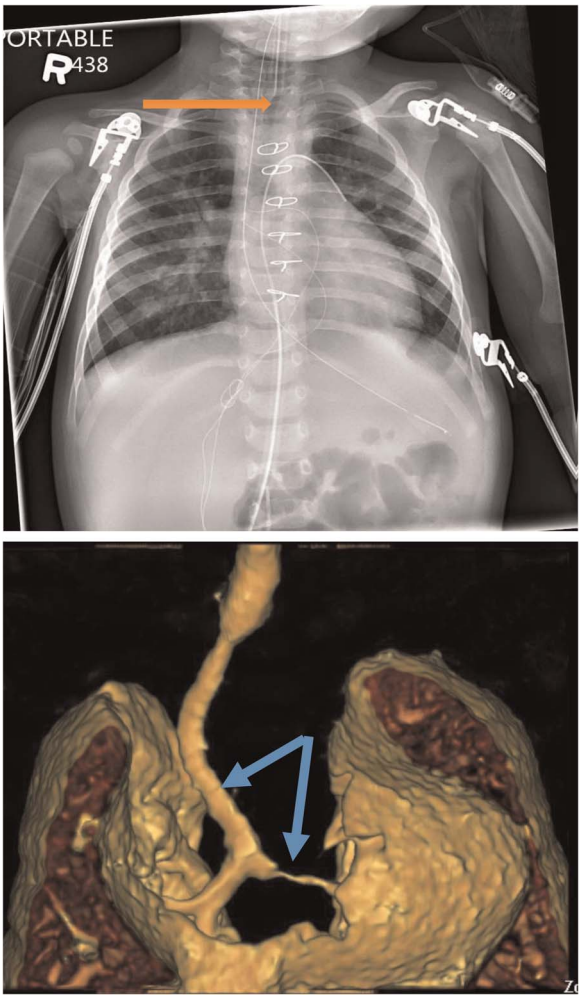

Figure 1.

P2029 - PERIOPERATIVE MANAGEMENT OF HYPERTENSION FOLLOWING TEH ROSS PROCEDURE IN PEDIATRICS A RETROSPECTIVE ANALYSIS

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Background: Perioperative hypertension following the Ross procedure is considered a potential complication that can lead to bleeding, autograft aneurysm and dilation. Tight blood pressure control can improve hemostasis; reduce the need for blood products and the need for reoperation.
Objectives and Methods: Study the correlation between blood pressure (BP) control in the immediate post cardiopulmonary bypass (CPB) period and postoperative bleeding, transfusion requirements and the length of ICU stay.

Results: See Tables 1–3 and Figures. This analysis includes 18 subjects. Demographic and clinical characteristics are presented in Table 1. There was no significant association between mean SBP and blood product exposure in the immediate post operative period. There was no significant association with the number of prior cardiac surgeries. In a linear regression analysis there was correlation between chest tube output and CPB duration ($r = 0.75$ for ages <2 years, $r = 0.55$ for ages > 2 years). In a univariate analysis there was no association between mean SBP at different time intervals in the postoperative period and chest tube output, but there was association between blood exposure and CPB duration ($P = 0.02$) Table 2, 3. The length of ICU stay was inversely related to the patient age ($p = 0.03$).
Conclusion: Our data demonstrated no significant association between bleeding and blood pressure control. There was correlation between CPB duration, CT output and transfusion requirements. Age was the only factor affecting the length of ICU stay

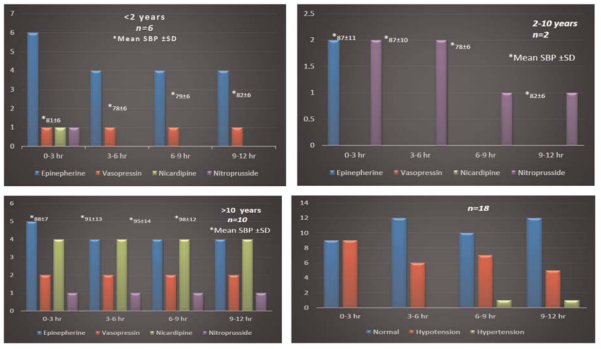

Figure 1.

Table 1. Demographic and Clinical Characteristics of Study Subjects.

Variable	All Patients (N = 18)
Age (yrs)	
0-2	6 (33%)
2-10	2 (11%)
>10	10 (56%)
Gender, M/F (%)	(67%)
Cardiac lesion, n (%)	
Critical AS/Bicuspid AV, Hypoplastic AV	14 (78%)
AR	1 (5%)
Combined AS, AR	3 (17%)
Shone Complex Diagnosis	5 (28%)
Previous Cardiac surgeries *(n%)*	8 (50%)
One previous cardiac surgery	4 (25%)
Two previous cardiac surgery	4 (25%)
Number of blood products exposure, Median Units (range)	
0-2 years	2 (1-4)
2-10 years	4 (0)
>10 years	7 (1-23)
Chest tube output first postoperative day, Median ± SD ml/kg/hr	0.48 ± 0.5
Cardiopulmonary Pump duration minute ± SD	292 ± SD
Length of ICU stay Days ± SD	
0-2 years	9 ± 5
2-10 years	3 ± 1
>10 years	4 ± 3
Length of Intubation Hours ± SD	
0-2 years	91 ± 60
2-10 years	15 ± 0.7
>10 years	20 ± 18
Return to OR for bleeding *(n%)*	2 (11%)

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Table 2. Univariate analysis of Blood product exposure and variables.

Covariate	Parameter estimate	P value	CI
Prior number of cardiac surgeries	2.78	0.07	(-0.25-5.83)
Cardiopulmonary Bypass time (min)	0.05	*0.02*	(0.01-0.09)
Age (years)	0.32	0.07	(-0.03,0.6)

Table 3. Univariate analysis of mean Systolic BP at different time interval and chest tube output.

Covariate	Parameter estimate	P value	CI
Mean SBP 3-6 Hrs	-0.02	0.07	(-0.04,0.002)
Mean SBP 6-9 Hrs	-0.02	*0.09*	(-0.036,0.003)
Mean SBP 9-12 Hrs	-0.02	0.06	(-0.04,0.001)

P2048 - REFRACTORY VENTRICULAR FIBRILLATION IN REDO CONGENITAL CARDIAC SURGERY KNOW THY PATIENT'S CORONARY

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Background: Ventricular Fibrillation (VF) in children with congenital heart disease is uncommon but could be an indicator of poor prognosis. VF during open heart surgeries is not uncommon but may be transient. However, refractory VF is rare even in the setting of open heart surgery. Survival to hospital discharge is still very low in "In hospital VF".
Methods: – Patients aged (–to–) underwent redo open heart surgery for repair of structural heart defects between year —— and – at KK women and children's hospital. 1 patient had refractory ventricular fibrillation that was not responding to conventional therapy including cardioversion.
Results: The right ventricle (RV) was lacerated while performing redo sternotomy and the patient was cannulated emergently via femoral route for CPB. After the repair was completed, the rhythm evolved into VF during rewarming but was recalcitrant and could not be corrected despite correction of metabolic parameters and multiple attempts at cardioversion. An on-table intracardiac hemodynamic assessment showed mild RV to pulmonary artery (PA) gradient across the conduit on low CPB flows. Review of the angiographic images to identify anatomical issues showed a small calibre left coronary system but normal anatomy. However, the left coronary artery patency could not be established beyond the bifurcation when probed. Hence, the RV –PA anastomosis was completely revised along with mobilization of the posterior adhesions,that resolved the VF, as the left coronary artery patency was re-established.
Conclusions: 1. Redo cardiac surgeries could compromise coronary circulation through adhesions even if the anatomical substrate does not have coronary anomalies.
2. Coronary hypo perfusion as an aetiology should always be sought in the context of prolonged refractory VF 3. Anatomical revision of the surgical repair may be required to mitigate refractory VF in some patients.

P2243 - CUSTODIOL FOR MYOCARDIAL PROTECTION FOR THE ARTERIAL SWITCH OPERATION RETROSPECTIVE ANALYSIS IN A SINGLE CENTER

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Background: Over 30% of the European pediatric cardiac surgery centers use cold Histidine-tryptophane-ketoglutarate (Custodiol) cardioplegia1. Neonatal procedures often imply aortic division, however repeated ostial canulation may cause intimal insult and affect long-term results. Custodiol cardioplegia is appealing since it has been suggested that a single-dose of cold Custodiol is safe for long aortic cross-clamping durations in infants2. Since cardiac troponins are specific markers of myocardial damage3, we thought investigate the association between myocardial no-flow duration and postoperative troponins in neonates undergoing the arterial switch operation with Custodiol cardioplegia.
Methods: 50 ml/kg Custodiol were administrated at 4 °C in the aortic root at cross-clamping, and repeated whenever judged necessary by the attending surgeon. High-sensitivity troponin was monitored postoperatively. The relationship between the longest myocardial no-flow duration per patient, i.e. the longest duration between two cardioplegia administrations and/or aortic unclamping, and the troponin concentration-time integral within 48 hours of surgery was analyzed using linear models stratified according to the coronary pattern.
Results: 101 patients were analyzed: age 6.0 ± 5.4 days, weight 3.7 ± 4.7 kg, cardiopulmonary bypass (CPB) duration 110.2 ± 53.2 min, cross clamping duration 75.3 ± 19.1 min, temperature during cross-clamping 31.2 ± 1.6 °C, duration of mechanical ventilation 5.9 ± 8.9 days, ICU stay 8.7 ± 9.5 days, 33

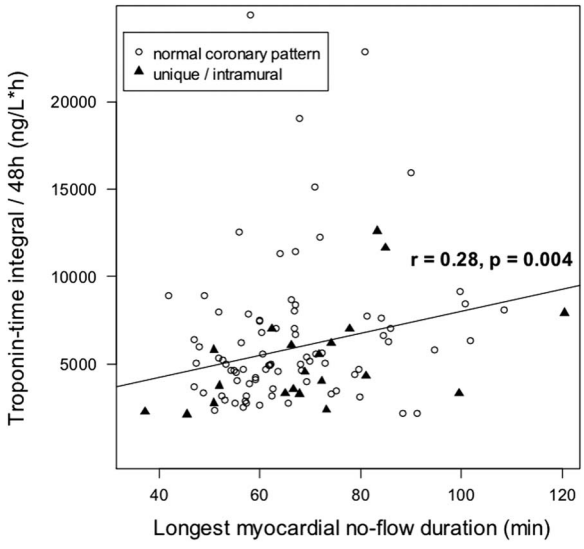

Figure.

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had delayed sternal closure, 1 required ECMO and died. The longest myocardial no-flow duration ranged 37-120 min, mean 67.2 ± 15.3 min, 26 patients had several cardioplegia administrations. Postoperative troponin peaked at 12565.3 ± 8994.8 ng/L. The longest myocardial no-flow duration was significantly associated with the troponin concentration-time integral ($R = 0.28$, $p = 0.004$), regardless of the coronary anatomy, the CPB duration and temperature, and with the need for delayed sternal closure ($p = 0.003$), but not with other outcome variables.
Conclusion: These results suggest that there is a risk of myocardial damage when single-dose Custodiol is used for long cross-clamping durations in neonates. 1.JECT 2012:186; 2.AFAR 2011: S17; 3.Eur Heart J 2000:1502.

P2400 - INCREASED LACTATE LOAD OF OLDER RED BLOOD CELL PREPARATIONS INCREASES BLOOD LACTATE CONCENTRATIONS IN INFANTS DURING CARDIAC SURGERY

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Background: The 2016 update of the red blood cell transfusion guidelines recommend the use of packed red blood cells (RRBC) selected at any point within their licensed dating period in all patients, including neonates1. Whereas the update included exclusively trials in premature neonates with reduced transfusion requirements, infants require massive transfusions during cardiopulmonary bypass (CPB). Older PRBCs contain high lactate concentrations, and there is evidence of slower lactate metabolism during CPB2. Since hyperlactatemia during CPB is considered to be an early and specific indicator of tissue hypoxia, it has become a major tool for hemodynamic monitoring and therapeutic guidance3. We thought investigate the relationship between PRBCs age and lactatemia during CPB in small infants.
Methods: During three month were explored the relationships between the PRBCs age, lactate concentration, and lactatemia on bypass in consecutive infants. Haematocrit was maintained >35% and venous saturation (SvO2) was maintained >70%. Data are shown as medians and interquartile ranges. Analyses used the Mann-Whitney test and linear regression.
Results: In 135 PRBCs, the PRBCs age (3-25days) correlated significantly with the lactate concentration (7.6-23 mmol/L), $r = 0.71$, $p < 0.01$. In 21 infants weighting <5 kg, aged 8 days [3-19], the prime volume was 218 mL [199-228], CPB duration was 169 min [128-274], CPB temperature was 32.8 °C [31.9-34.2], the duration of SvO2 excursions below 70% per patient was 0.5 min [0-3.4]. When compared with patients having received PRBCs <5days, patients having received PRBCs ≥5days had higher lactate concentrations in the prime 2.5 mmol/L [2-2.7] vs 1.6 mmol/L [1.5-1.7], $p = 0.04$ and at the end of CPB, 5.4 mmol/L [4.4-6.5] vs 2.7 mmol/L [1.8-2.8], $p < 0.001$, regardless of CPB duration, temperature, estimated patient blood volume or duration of SvO2 excursions below 70%.
Conclusion: When administered during CPB in small infants, PRBCs ≥5days represent a potential source of hyperlactatemia, and could lead to unnecessary and/or deleterious therapeutic interventions. 1. JAMA,2016:2025; 2. ICM,2003:1279; 3. ICM,2005:98

P2484 - CLINICAL OUTCOMES WITH THE USE OF LEVOSIMENDAN IN PATIENTS WITH HYPOPLASTIC LEFT HEART SYNDROME BROUGHT TO NORWOOD PROCEDURE

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Introduction: Hypoplastic left heart syndrome (HLHS) can cause approximately 23% of all cardiac deaths in the first week of life and 15% in the first month, making it a strong morbidity and mortality indicator in specialized centers. Levosimendan, due to its inodilatory characteristics without increasing oxygen consumption, has been widely used in the adult population in high risk patients for cardiac surgery. Even though its use in children has been limited, given the physiological characteristics of HLHS, levosimendan arises as a possible inotropic alternative in the management of these patients.
Materials and Methods: Retrospective and descriptive study in a fourth level center in Colombia. Inotropic management (milrinone + adrenaline) was compared with (Levosimendan + adrenaline), initiated in the perioperative period. 30-day mortality and hospital discharge, as well as the clinical outcomes given by length of stay in the ICU, hospital stay, mechanical ventilation time, neurological compromise, need for ECMO, and renal failure were analyzed.
Results: Between January 2009 and December 2015, 55 patients diagnosed with HLHS were taken to Norwood procedure with pulmonary systemic shunt or RV-PA shunt, 17 (31%) patients received levosimendan and 38 (69%) received milrinone (Table 1). When comparing the two managements, the levosimendan group had an in-hospital mortality of 17.7% Vs 50% (p 0.036) and a decrease in the presence of postoperative renal failure 23.5% vs. 54.1% (p 0.036). No differences were found in 30-day mortality (p0.303) and in the clinical outcomes analyzed (Table 2).
Conclusions: In our study, levosimendan appears to be an alternative for inodilator management in the peri and postoperative period of HLHS patients undergoing Norwood procedure because it was found to significantly reduce the presence of postoperative renal failure and mortality.

Table 1.

	With Levosimendan	Without Levosimendan	*P*
In-hospital mortality	19 (50%)	3 (17.7%)	*0,036*
Renal failure	20 (54.1%)	4 (23.5%)	*0,036*
30 days Mortality	11 (29%)	2 (11.8%)	*0,303*
POP stay (days)	68 (23-180)	73 (6-171)	*0,678*
POPPICU stay (days)	24 (1-126)	21 (1-167)	*0,654*
Neurological impairment	14 (37.8%)	6 (35.3%)	*0,857*
ECMO	17 (44.7%)	8 (47.1%)	*0,551*
Inotropic days	17 (1-116)	12 (1-136)	*0,222*
Mechanical ventilation (days)	11.5 (1-118)	10 (1-75)	*0,523*

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Table 2.

Total patients	55		Without Levosimendan	With Levosimendan	P
Levosimendan	17	31%			
Currently alive	20	36%	12 (31.6%)	8 (47.1%)	*0,27*
Age income					*0,11*
Median	#iREF!		1,5	5	
Rank	0-38		0-38	0-19	
Gender (male)	38	69%	26 (68.4%)	12 (70.6%)	*0,87*
Weight at surgery					*0,051*
Median	3370		3325	3700	
Rank	2500-4800		2500-4800	2900-4800	
Phenotype		0%			*1*
1. AA/AM	23	42%	16 (42.1%)	7 (41.2%)	
2. AA/EM	12	22%	8 (21.1%)	4 (23.5%)	
3. EA/EM	15	27%	10 (26.3%)	5 (29.4%)	
4. EA/AM	5	9%	4 (10.5%)	1 (5.9%)	

P2509 - ROLE OF ROTEM IN BLOOD PRODUCT TRANSFUSION IN NEONATE ON ECMO UNDERGOING CONGENITAL DIAPHRAGMATIC HERNIA REPAIR

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Congenital diaphragmatic hernia (CDH) is a rare congenital anomaly. Due to the hypoplastic lungs and severe pulmonary hypertension, frequent usage of extracorporeal membrane oxygenation (ECMO) is deemed essential for sustenance of the neonate. However, during surgical intervention, life threatening bleeding can occur in such patients. We present a case report of a neonate who underwent CDH repair with undergoing ECMO support, emphasizing the role of rotational thromboelastometry (ROTEM) in bed side checking coagulation profile and facilitating transfusion of blood products immediately.

Methods: The neonate was antenatally diagnosed having isolated CDH. He was delivered by normal vaginal delivery and was put on ECMO on day two after birth. Primary repair of CDH was planned in neonatal intensive care unit as no improvement was reported in physiological condition, with persistent poor lung to head ratio despite being on 100% oxygen.

Results: The neonate was on constant heparin infusion to maintain ECMO. Intraoperatively ROTEM system was used to check coagulation level effectively in rapidly changing haemodynamic status. Blood products were transfused according to the ROTEM result interpretation. The coagulation profile was monitored intermittently by the laboratory to validate ROTEM results. Total of more than seven hundred millilitre of blood products were transfused in a 2.96 kilogram neonate intraoperatively during 6 hours of operation.

Conclusion: This is the first neonatal non-cardiac surgery to the best of our knowledge where ROTEM was utilized to efficiently manage the coagulation profile in a neonate with so much amount of blood products transfused intraoperatively for CDH repair. The perioperative coagulation parameters remained in accordance with the laboratory results. Disclosure of Interest: None declared.

P2524 - EARLY SURGICAL OUTCOME OF PEDIATRIC TETROLOGY OF FALLOT SURGERY IN INDIAN POPULATION; A SINGLE CENTRE ANALYSIS

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Background: Tetrology of fallot (TOF) is the most common cyanotic congenital heart disease. Management of total surgical correction of the tetrology patients remains challenging during the perioperative period in developing countries. Variability in early surgical outcome in tetrology patients in developing countries could be because of late presentation, surgical factors, associated anomalies, infections and economic factors. The aim of this study was to determine the early in-hospital mortality after TOF surgery and to identify the factors associated with adverse outcome in the early postoperative period.

Methods: This retrospective study involved 207 TOF patients who underwent surgery over a period of two years in a tertiary hospital in India.Age, sex, demographics, type of surgery, additional intraoperative echo findings, early postoperative mortality, duration of mechanical ventilation, duration of ICU stay and postoperative complicationswere recorded and analysed.

Results: In our centre, TOF surgical patients represented 21.75% of overall paediatric congenital heart surgery. Among the TOF patients who underwent surgery, two-third of patients were male. The mean age for shunt surgery and total intracardiac repair were 8 months and 7 years respectively. The overall surgical mortality in TOF patients was 10.6%. While mortality that occurred in shunt surgeries was 27.27%, total intracardiac repair patients had 7.4% early in-hospital mortality. Non-survivors in total intracardiac repair patients had higher aortic cross clamp time, bypass time and prolonged intensive care stay. Major aorto-pulmonary collaterals were present in 6% of patients. Junctional ectopic tachycardia (JET) remains the most common complication in the post operative period.

Conclusion: Late presentation for surgery, shunt surgeries in neonatal period, prolonged bypass time, prolonged cross clamp time and prolonged ICU stay were the factors associated with higher early surgical mortality in TOF patients in the Indian subcontinent.

P2536 - BENEFITS OF USING MICROCUFFED TRACHEAL TUBE IN PATIENTS UNDERGOING CONGENITAL CARDIAC SURGERIES EVIDENCE FROM A PROSPECTIVE RANDOMIZED STUDY

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Background: It has been suggested that compared to conventional uncuffed tubes, high volume-low pressure microcuffed tracheal tubes in smaller children are beneficial. However, there is paucity of evidence of its usage in children undergoing congenital cardiac surgery. Aim of this study was to assess if microcuffed tracheal tubes in neonates and younger children undergoing congenital cardiac surgery is associated with better outcomes than uncuffed tubes.

Methods: We carried out this single centre, prospective, double blinded randomized study between June and November 2016. 80 patients were randomized into those receiving microcuffed tracheal tubes and conventional uncuffed tubes. Primary outcome

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was post-extubation stridor. Secondary outcomes measured included number of tube changes, volume of anaesthetic gases required and cost incurred.
Results: The 2 groups were comparable in terms of baseline characteristics and duration of intubation.Incidence of stridor was significantly higher in conventional uncuffed tubes (12(30%) vs 4 (10%), $p = 0.04$) and so was the the number of tube changes required (17/40 (42.5%) vs. 2/40(5%); $p = < 0.001$). Tube change was associated with more than three-fold risk of stridor (odds ratio 3.92 (95% CI 1.23-12.43). Isoflurane (29.14 ± 7.01 ml vs 19.2 ± 4.81 ml; p value < 0.0001) and Oxygen flow requirement (p value <0.0001) and the resultant cost (p value <0.0001) were all significantly higher in the conventional uncuffed group.
Conclusion: Microcuffed paediatric tracheal tube is associated with significantly lower incidence of stridor, tube changes and anaesthetic gas requirement. This leads to significant cost-reduction that offsets the higher costs associated with usage of microcuffed tracheal tube.

P2750 - MULTIDISCIPLINARY IMPLEMENTATION OF EVIDENCED BASED GUIDELINES FOR EXTUBATION IN THE OPERATING ROOM FOLLOWING PEDIATRIC CARDIAC SURGERY

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Background: Extubation in the operating room (OR) following pediatric cardiac surgery has become commonplace in contemporary practice. Its benefits however must be measured against its risks, most notably of which is extubation failure. We conducted an evidence-based quality improvement initiative at our institution aiming to minimize complications associated with extubation in the OR in this patient population.
Methods: A multidisciplinary team created the following guidelines for extubation in the OR after pediatric cardiac surgery (based on current literature): age >1 month; STAT mortality category ≤4; no known airway anomaly; cardiopulmonary bypass duration <150 minutes; non-emergent procedure; no significant intraoperative bleeding or respiratory complications; and postoperative beta-agonist therapy limited to dopamine infusion ≤5 mcg/kg/min (based on institutional preferences). A quality improvement initiative using these guidelines was conducted between 1/2015-12/2016, and data from this period were compared to historical data from 1/2011-12/2014.
Results: From 2011-2014, we extubated 953 of 1989 patients in the OR (48%), of which 13 failed extubation (1.4%). In 2015-2016, we extubated 317 of 960 patients in the OR (33%), of which 3 failed extubation (0.9%). The observed decrease in the proportion of patients extubated in OR occurred primarily in STAT category 1 and 2 procedures (Table). In this subset, median duration of postoperative mechanical ventilation increased slightly, from 0 hours (25%,75%: 0-7 hours) to 1.7 hours (25%,75%: 0-24 hours).
Conclusions: During our evidence-based quality improvement initiative in children undergoing cardiac surgery, we minimized the occurrence rate of extubation failure in the ICU following extubation in the OR. The clinical significance of the concomitant decrease in OR extubation rate and increase in duration of mechanical ventilation associated with this initiative is unclear. Further work is needed to optimize the practice of extubation in the OR after pediatric cardiac surgery.

Table. Proportion of Patients Extubated in the Operating Room.

STAT Mortality Categories	2011-2014	2015-2016
1	360/480 (75%)	122/208 (59%)
2	288/523 (55%)	80/241 (33%)
3	60/197 (30%)	27/99 (27%)
4	42/270 (16%)	15/153(10%)
5	2/59 (3.3%)	1/28 (4%)

P2768 - IMPACT OF ANESTHETIC MANAGEMENT DURING DIAGNOSTIC CATHETERIZATIONS PERFORMED IN SINGLE VENTRICLE PATIENTS PRIOR TO STAGE II PALLIATION

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Background: Diagnostic cardiac catheterization plays a pivotal role in the assessment of a single ventricle (SV) infant's physiologic status prior to stage II palliation and wide variability in practice patterns exists. We sought to compare the safety and efficacy of sedation management strategies during the pre-stage II catheterization.
Methods: We performed a single center retrospective cohort study of SV infants undergoing pre-stage II catheterization between 2010-2015. Safe anesthesia was defined by absence of cardiac or respiratory events within 24 hours of the procedure and no need for post-catheterization ICU admission. Effective anesthesia was defined by stable hemodynamics (<2 episodes of heart rate increase requiring anesthetic boluses) and no need to convert from sedation to general anesthesia. Multivariate logistic regression models were utilized to compare results.
Results: Among 90 SV infants sedated for diagnostic pre-stage II catheterization, the anesthetic was safe in 72 (80%) and effective in 49 (54%), while 8 (89%) required conversion to general anesthesia. Demographics and perioperative variables are detailed in Table 1. All infants received dexmedetomidine infusions and 96% received midazolam. Fentanyl and ketamine were the next most commonly employed medications with less frequent boluses of propofol, morphine, and diphenhydramine. A safe anesthetic was independently associated with higher baseline weight and baseline Sa02, while negatively associated with cardiologist #2 (Table 2). An effective anesthetic was independently associated with higher weight (Table 3). There were no differences in remaining baseline demographics, procedural duration, cardiologists or anesthesiologist years of experience. Safe and effective anesthetic management was associated with a ketamine loading dose of >0.5 mg/kg and infusion, and independently associated with higher weight, while negatively associated with cardiologist #2 (Table 4).
Conclusions: Safe and effective sedation can be performed on SV infants undergoing pre-stage II catheterization with utilization of anesthetic medications that do not depress respiration.

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Table 1. Demographic and Clinical Characteristics of Sedation Subjects.

Characteristics	n (%)
Male	57 (63.3)
Age (wk)	21.2 ± 5.5
Weight (kg)	6.0 ± 1.0
Pre-Operative	
Baseline SpO_2	77.9 ± 6.6
Intra-Operative	
Lowest pH	7.36 ± 0.05
Highest pCO_2	43.7 ± 6.05
Procedural duration (min)	83 ± 30.7
Failed sedation	8 (9.9)
Post-Operative Disposition	
Monitored ward	50 (55.6)
Intensive care unit	2 (2.2)
Same day discharge	36 (40)

Abbreviations: SpO_2, oxygen saturation.
Data presented as n (%) or mean ± standard deviation.

Table 2. Odds of Safe Anesthetic during Diagnostic Cardiac Catheterization.

	Unadjusted			Adjusted		
Variables	**OR**	**95% CI**	p^a	**AOR**	**95% CI**	p^b
Weight, kg	1.87	1.00,3.49	0.05	2.14	1.04,4.38	0.038
Baseline SaO_2	1.10	1.01 1.20	0.029	1.12	1.01,1.23	0.030
Cardiologist#2	0.20	0.64,0.63	0.006	0.133	0.04,0.52	0.004

AOR = adjusted odds ratio, CI = confidence interval, OR = unadjusted odds ratio. [a]*p*-value from univariate logistic regression. [b]*p*-value from multivariable logistic regression.

Table 3. Odds of Effective Anesthetic during Diagnostic Cardiac Catheterization.

	Unadjusted			Adjusted		
Variables	**OR**	**95% CI**	p^a	**AOR**	**95% CI**	p^b
Weight, kg	1.80	1.12,2.88	0.014	1.89	4.15,3.09	0.012
Baseline SaO_2	0.99	0.93,1.06	0.803			
Ketamine Load >0.5 mg/kg and ketamine infusion	4.39	0.89,21.61	0.069			

AOR = adjusted odds ratio, CI = confidence interval, OR = unadjusted odds ratio.
[a]*p*-value from univariate logistic regression. [b]*p*-value from multivariable logistic regression.

Table 4. Odds of Safe and Effective Anesthetic during Diagnostic Cardiac Catheterization

	Unadjusted			Adjusted		
Variables	**OR**	**95% CI**	p^a	**AOR**	**95% CI**	p^b
Weight, kg	2.07	1.27,3.38	0.004	2.44	1.41,4.20	0.001
Baseline SaO_2	1.01	0.95,1.08	0.75			
Cardiologist #2	0.31	0.10,0.95	0.041	0.26	1.41,4 20	0.043
Ketamine Load >0.5 mg/kg and ketamine infusion	5.38	1.09,26.49	0.039	5.56	0 99,31.09	0.051

AOR = adjusted odds ratio, CI = confidence interval, OR = unadjusted odds ratio.
[a]*p*-value from univariate logistic regression. [b]*p*-value from multivariate logistic regression.

P2831 - EFFECT OF AGE OF BLOOD AND PRIME LACTATE LEVEL ON THE PERIOPERATIVE OUTCOME IN CHILDREN UNDERGOING COMPLEX CONGENITAL HEART SURGERY

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Aim: To Study The Effect of Age of Blood and Prime Lactate on the Perioperative Outcome in Children Undergoing Complex Congenital Cardiac Surgery.
Methods and Materials: Sri Ramachandra Medical Centre ethical committee permission was obtained before the study. Patients who were undergoing Complex Congenital Heart Disease From January 2012 to December 2013, 54 were retrospectively selected for this study. These 54 patients were divided into 2 groups depending upon the age of the blood they were subdivided into Group A(≤3 days) and Group B (>3 days) and depending upon the prime lactate values. Group I (≤10 mmol/l) and Group II (>10 mmol/l).
Inclusion Criteria: Patients of either sex, Patient age ≤10 years, patients with complex congenital cardiac disease.
Exclusion Criteria: Patients age >10 years and patients who had plasma as prime.
Conclusion: The storage time of RBCs deleteriously determine the postoperative morbidity of patients in terms of renal parameters. The increase in the prime lactate also significantly contributes to the elevated renal parameters and bilirubin with poor neurological outcome. This study has proven it significantly. Hence, the use of the freshest possible blood is suggested for CPB priming and maintaining the prime lactate as low as possible to avoid morbidity.

P2870 - SURGICAL OUTCOME OF THE VENTRICULAR SEPTAL DEFECT REPAIR WITH SEVERE PULMONARY HYPERTENSION IN TERTIARY CARE HOSPITAL IN INDIA

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Background: Surgical closure of ventricular septal defect (VSD) is the most common acyanotic congenital heart surgery in pediatric patients. The nature of complications and outcomes in VSD patients with severe pulmonary hypertension (PH) undergoing surgery varies in developing and developed countries. The main purpose of this study was to determine the early outcome in VSD patients with severe PH during perioperative period.
Methods: This retrospective review included 35 pediatric patients of VSD with severe PH that were surgically treated for correction between January 2016 and December 2016. Age at time of surgery, sex, type of defect, pulmonary arterial pressure, shunt direction, ratio of blood flow in pulmonary circulation to systemic circulation (QP; QS), duration of mechanical ventilation, and duration of stay in the intensive care unit (ICU), complications and mortality during perioperative period were noted and analyzed.
Results: VSD patients with severe PH constituted 22.5% of overall VSD with equal gender distribution. The overall mortality occurred in cohort was 8.57%. The median age at which patients were operated was 3.5 years. The average mechanical ventilation and intensive care stay were 12 hours (IQR: 2–98) and 5 days (IQR: 1-18), respectively. Of the 35 patients, 3 patients had bidirectional shunt, which was incidentally diagnosed during

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perioperative period with all three patients had QP:QS ratio more than two. Among the three patients with bidirectional shunt, one patient developed congestive cardiac failure in the immediate post operative period. Overall, 14.5% had CCF during perioperative period.
Conclusion: We concluded that VSD with severe PH continues to be a major cause of postoperative morbidity and have increased the duration of mechanical ventilation and ICU stay in these patients in developing countries. Advances in the management of PH during perioperative period drastically reduced the mortality in these patients over the past few years.

P2918 - THE USE OF SOMATOSENSORY EVOKED POTENTIAL (SSEP) AND ELECTROMYOGRAPHY (EMG) MONITORING TO DESCRIBE BRACHIAL PLEXUS AND FEMORAL NERVE CHANGES DURING CARDIAC CATHETERIZATION PROCEDURES IN CHILDREN

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Introduction: Children with congenital heart disease frequently require cardiac catheterization procedures and common to almost all procedures are the need for femoral vessel access and positioning the arms above the head. These procedures in children often last a long time, typically 4 to 6 hours, due to the complexity of their congenital heart disease and the difficulties in obtaining vascular access. Thus, many children undergoing cardiac catheterization require general anesthesia to tolerate the procedure and remain immobile, so optimal images and data can be obtained. Peripheral nerve injury may mar an otherwise successful cardiac catheterization and result in a potentially disabling injury for the patient and litigation involving the health care team. Examination of the American Society of Anesthesiologists Closed Claims Database showed that 16% of all claims in adults were related to nerve injury. The most frequent sites of injury were the ulnar nerve (28%) and brachial plexus (20%). Many patients with congenital heart disease now survive to adulthood and this patient population may be at particular risk of brachial plexus injury during cardiac catheterization procedures. There are several reports of brachial plexus injury during cardiac catheterization in children but no reports of using SSEP monitoring to detect brachial plexus changes or EMG to record inadvertent femoral nerve puncture during a cardiac catheterization procedure.
Study Aims: This single center prospective study aims to:

- Describe changes in the SSEPs in the brachial plexus during cardiac catheterization procedures
- Describe changes in EMG activity recorded over the quadriceps related to inadvertent femoral nerve puncture
- Determine if SSEP changes are clinically important for patients and if monitoring technology may be a useful tool to prevent nerve injury in patients.

Results: 42 children were successfully enrolled into the study and the data will be presented for the first time at WCPCCS.

P2930 - "ON THE TABLE" EXTUBATION IMMEDIATELY AFTER PEDIATRIC AND CONGENITAL HEART SURGERY

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Background: "On the Table" extubation immediately after pediatric and congenital heart surgery ("immediate extubation" [IE]) has been described as safe and cost effective in adult patients undergoing open heart surgery. Early extubation (EE), or extubation within 6 hours after the operation, has shown a decrease in postoperative length of stay in patients having Tetralogy of Fallot repair and the Fontan operation. We reviewed our experience with IE after pediatric and congenital heart surgery, comparing our rates of IE to the national aggregate rates documented in the Society of Thoracic Surgeons Congenital Heart Database (STS CHD).
Method: We performed a retrospective review of all patients who underwent open heart surgery at a single institution between January 2010 and December 2016. Rates of IE were calculated for our patients stratified by age groups and STAT Mortality Categories, and these rates were compared to national aggregate data.
Results: Table 1 documents rates of IE stratified by age groups and STAT Mortality Categories, for our patients and for national aggregate data.
Conclusion: IE after pediatric and congenital heart surgery can be accomplished safely in selected patients across all ages, from neonates (sparingly) through adulthood (usually), and across all STAT Categories, although only sparingly for STAT Category 5. This strategy represents an opportunity for reducing postoperative length of stay and cost.

Table 1. Immediate extubation(IE) rates in the JHACH cohort (N = 1528) and comparison with the entire STS cohort (N = 60394).

Characteristic	JHACH	Entire STS
Extubation		
IE	1015/1528 (66.6%)	12099/60394 (20%)
IE according to age		
Neonates	47/294 (15.9%)	417/13918 (3.0%)
Infants	360/523 (68.8%)	2644/20378 (13.0%)
Children	510/584 (87.3%)	7758/22468 (34.5%)
Adults	98/122 (80.3%)	1280/3629 (35.2%)
IE according to STAT		
1	368/419 (87.8%)	5177/15296 (33.8%)
2	467/564 (82.8%)	4861/19236 (23.3%)
3	51/116 (43.9%)	780/6280 (12.4%)
4	97/296 (32.8%)	787/11706 (6.7%)
5	4/93 (4.3%)	97/2373 (4.1%)

ELECTROPHYSIOLOGY

P1020 - LATE POST OPERATIVE ARRYTHMIA AT TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION

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Introduction: The frequency of events of anomalous pulmonary venous connection (APVC) varies in the different series from 5.9 to 7.1 / 100 000 live births, representing 1 to 5% of cases of congenital heart disease. Since the current care has develope, the need to approach possible late complications, such as arrhythmias, is important.
Objective: To evaluate the distribution of arrhythmias and possible related variables in patients in the late postoperative APVC.
Method: Medical record review and clinical and noninvasive arrhythmia evaluation were used for data analysis of 20 patients who underwent isolated APVC. Statistical analysis for the detection of related variables.
Results: Clinical and echocardiographic evaluations showed no significant changes. Of the 20 patients, 13 (65%) showed some abnormalities for age according to the 12-led EGC. By Holter, six patients (30%) presented atrial and/or ventricular ectopy with higher frequency than mild. Junctional rhythm and 2: 1 atrioventricular occurred in one patient (5%). Analyzing the criteria for sinus node dysfunction, bradycardia was observed by Holter in 30% of the patients. There were no pauses longer than two seconds. By Ergometric test, 82% of children had chronotropic deficit. Follow-up more than 60 months and abnormal ECG were related to the presence of arrhythmias.
Conclusion: The long term follow-up of patients undergoing anomalous pulmonary venous connection showed that regardless of the good clinical and echocardiographic results, the possibility of occurrence of atrial and/or ventricular arrhythmias highlight attention to the need for maintenance of regular clinical evaluations.

P1023 - A GIANT LEFT VENTRICLE ANEURYSM IN A 12 YEAR OLD MALE CHILD

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Left ventricle aneurysm is a rare finding in children. We report a case of a 12 year old male who was brought to the emergency room for non-sustained ventricular tachycardia. He was previously diagnosed with Rheumatic Heart Disease but additional imaging studies (Transthoracic Echocardiogram and Chest CT scan) revealed a giant LV aneurysm. The patient was started on anticoagulation therapy due to the risk of thromboemboli and was given medication for left ventricular systolic dysfunction, in preparation for elective surgery. An endoventricular repair technique under cardiopulmonary bypass was done to repair the LV after removing the aneurysm. This was noted to be adherent to pericardium with neovascularization. An intraoperative transesophageal echocardiogram showed significant decrease in LV end diastolic dimension size from 6 cm to 4.8 cm, with moderate mitral regurgitation, sinus rhythm, with good LV contractility post repair. His post-operative rhythm was sinus, with no recurrence of VT. Histologic examination revealed a non-specific inflammatory process. Immediate postoperative course was uneventful and our patient was discharged after four days. On follow-up, a repeat TTE showed improved LV contractility with mild mitral regurgitation and remained sinus rhythm on ECG. Patients with a LV aneurysm are at potential risk for life-threatening ventricular tachyarrhythmias which may sometimes occur as the first clinical presentation. This can be reliably diagnosed with modern diagnostic imaging. The etiology of which is still mostly infectious in children. Successful surgical repair using standard operative techniques can be done which render excellent post-operative outcomes.
Key words: LV aneurysm, endoventricular repair, ventricular tachycardia

P1026 - RATE CONTROL BY TRANSOESOPHAGEAL ATRIAL QVERDRIVE PACING FOR REFRACTORY SUPRAVENTRICULAR TACHYCARDIA WITH SEVERE VENTRICULAR DYSFUNCTION – A BRIDGE TO RECOVERY

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Introduction: Tachyarrhythmia-induced cardiomyopathy (TIC) is a rare yet life-threatening phenomenon in children. TIC has been defined as myocardial dysfunction that is entirely or partially reversible after control of the responsible tachyarrhythmia and is typically caused by an incessant supraventricular tachycardia. In cases of unsuccessful termination of the tachycardia cardiogenic shock may occur. Several authors favour the use of mechanical circulatory support in such cases. In view of these partly severe cases the authors would like to present the option of transoesophageal overdrive pacing (TOP) as a tool for initial heart rate control and hemodynamic stabilization of patients with TIC refractory to conventional strategies of tachycardia termination.
Methods: Chart review for patients receiving TOP during the last 5 years in two centers was performed.
Results: A case series of successful TOP including technique and outcomes is depicted.
Conclusion: Rate control through TOP is a safe and effective option to restore acceptable hemodynamics in infants with refractory supraventricular tachycardia and severe impairment of ventricular function. It presents the possibility of immediate heart rate control and offers time for myocardial recovery and safe implementation of antiarrhythmic drug therapy.

P1088 - MEASURES OF HEART RATE VARIABILITY DEPEND ON AGE OF HEALTHY CHILDRENINTRODUCTION HEART RATE VARIABILITY (HRV) IS THE PHYSIOLOGICAL PHENOMENON CAUSED BY CHANGES OF DURATION BETWEEN CONSECUTIVE CARDIAC CYCLES OF SINUS ORIGIN. HRV HAS A PROVEN CLINICAL AND PHYSIOLOGICAL VALUE IN ADULTS FOR EXAMPLE FOR THE DIAGNOSIS OF DIABETIC AUTONOMIC NEUROPATHY SELECTION OF HIGH RISK POST INFARCTION PATIENTS OR PEOPLE UNDER INCREASED EMOTIONAL

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Heart rate variability (HRV) is the physiological phenomenon caused by changes of duration between consecutive cardiac cycles of sinus origin. HRV has a proven clinical and physiological value in adults, for example for the diagnosis of diabetic autonomic neuropathy, selection of high-risk post-infarction patients or people under increased emotional stress. In general, many HRV parameters become reduced with advancing age in adults, however data on the association between age and HRV are sparse in children. In this study we examined the correlation between the age of healthy children and selected measures of HRV in 24-hour ECG Holter recordings.
Material and methods: One hundred healthy children (51 girls), aged 3-18 years old, underwent 24-hour ECG Holter recording (Schiller Medilog Darwin, Switzeland) . After a detailed analysis, all cardiac cycles (RR intervals) with an annotations about the beat type were exported and analyzed with in-house software with the use of Lomb periodograms and the Poincaré plots analyses. The association between age and HRV parameters was analyzed with the nonparametric Spearman test.
Results: There were significant and positive correlations between children's age and the mean duration of RR interval ($r = 0.55$; $p < 0.0001$), the power of ultralow (ULF; $r = 0.42$; $p < 0.0001$), very low (VLF; $r = 0.35$; $p = 0.0004$) and low frequency (LF; $r = 0.3$; $p = 0.0028$), a measure of total HRV (SDNN; $r = 0.33$; $p = 0.0008$) and long-term HRV (SD2; $r = 0.37$; $p = 0.0002$). Neither the power of high frequency (HF) nor the measure of short-term HRV (SD1) were significantly correlated with age.
Conclusion: We show that most HRV measures derived from the 24-hour ECGs are significantly related to the age of healthy children. HRV indices which describe ultralow, very low and low frequency oscillations or total and long-term variability increase with age while HRV parameters related to either high frequency or short-term HRV are not dependent on age. The physiological and clinical value of our findings requires.

P1089 - HEART RATE ASYMMETRY IS PRESENT IN HEALTHY CHILDREN.INTRODUCTION HEART RATE ASYMMETRY (HRA) IS THE PHYSIOLOGICAL PHENOMENON CAUSED BY AN UNEQUAL CONTRIBUTION OF HEART RATE DECELERATIONS AND ACCELERATIONS TO DIFFERENT FEATURES OF HEART RATE VARIABILITY AND STRUCTURE. SO FAR HRA HAS BEEN DESCRIBED IN ECGS OF DIFFERENT LENGTH FROM 1 MINUTE TO 24 HOUR ONLY IN ADULTS BUT NOT IN CHILDREN. IN THIS

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Heart rate asymmetry (HRA) is the physiological phenomenon caused by an unequal contribution of heart rate decelerations and accelerations to different features of heart rate variability and structure. So far HRA has been described in ECGs of different length from 1-minute to 24-hour only in adults but not in children. In this study we examined whether HRA is present in healthy children in 24-hour ECG Holter recordings.
Material and Methods: One hundred healthy children (51 girls), aged 3-18 years old, underwent 24-hour ECG Holter recording (Schiller Medilog Darwin, Switzeland). After a detailed analysis, all cardiac cycles (RR intervals) with annotations about the beat type were exported and analyzed with in-house software to quantify the expression of various forms of HRA. To quantify the contribution of heart rate decelerations (d) the following parameters were applied: C1d for the short-term, C2d for the long-term, and CTd for the total HRA. Additionally, the departure of the proportions of different forms of HRA from 0.5 was analyzed with the binomial test. Expression of HRA is shown as mean + /- SD while proportions as a part of 1 and its 95% confidence interval (CI).
Results: Mean expression of C1d was 55.4 + /-4.1%, of C2d 47.2 + /-3.0% and CTd 47.7 + /-2.5% - these results show that heart rate decelerations have a larger contribution to the short-term (C1d) and smaller to the long-term (C2d) and total (CTd) heart rate variability. At the same time, the proportion of children with $C1d > 50\%$ was 0.94 (0.87-0.98; $p < 0.0001$), $C2d < 50\%$ was 0.86 (0.78-0.92; $p < 0.0001$) and CTd 0.86 (0.78-0.92; $p < 0.0001$) – these results show that different features of HRA are present in the majority of healthy children.
Conclusion: We show that short-term, long-term and total heart rate variability has asymmetrical features in 24-hour ECGs recorded in healthy children. This means that physiological phenomenon of heart rate asymmetry is present children.

P1119 - ARRHYTHMIAS IN ADULTS WITH CONGENITAL HEART DISEASE WHAT ARE RISK FACTORS FOR SPECIFIC ARRHYTHMIAS

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Background: An increasing number of patients with congenital heart disease are now surviving into adulthood. This has led to the emergence of complications from the underlying congenital heart disease, related surgical interventions, and associated combordities. While the prevalence of particular arrhythmias with specific congenital heart disease has been previously described, a detailed analysis of all lesions and a large number of comorbidities has not been previously published.
Materials and Methods: Admissions with congenital heart disease were identified in the National Inpatient Sample. Associated comorbidities were identified. Univariate analysis was done to compare those risk factors associated with specific arrhythmias in the setting of congenital heart disease. Next, regression analysis was done to identify what patient characteristics and comorbidities were associated with specific arrhythmias.
Results: A total of 52,725,227 admissions were included. Of these, 109,168 (0.21%) had congenital heart disease. Of those with congenital heart disease, 27,088 (25%) had an arrhythmia. The most common arrhythmia was atrial fibrillation which was noted in 86% of those with arrhythmia followed by atrial flutter which was noted in 20% with congenital heart disease. The largest burden of arrhythmia was found to be in those with tricuspid atresia with a 51% prevalence of arrhythmia, followed by Ebstein anomaly which had an arrhythmia prevalence of 39%. Increasing age, male gender, double outlet right ventricle, atrioventricular septal defect, heart failure, obstructive sleep apnea, transposition of the great arteries, congenitally corrected transposition, and tetralogy of Fallot were frequently noted to be independent risk factors for specific arrhythmias.

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Conclusion: Approximately 25% of adult admissions with congenital heart disease are associated with arrhythmia. The burden of arrhythmia varies by the specific lesion and other risk factors as well. Understanding of these can help in risk stratification and can help devise strategies to lower this risk.

P1159 - PRENATAL DIAGNOSIS OF FETAL JUNCTIONAL ECTOPIC TACHYCARDIA USING MAGNETOCARDIOGRAPHY

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Background: Fetal tachyarrhythmia, characterized by a sustained fetal heart rate (FHR) above 180 bpm may be poorly tolerated and cause ventricular dysfunction leading to heart failure and hydrops, and ultimately to fetal demise. We present the first case of fetal junctional ectopic tachycardia (JET) diagnosed in utero using fetal magnetocardiography (fMCG) and not associated with maternal SSA/SSB antibodies.
Methods: N/A
Results: The patient was first referred to us at 20 weeks gestation for non-variable fetal tachycardia (FHR 180 to 190 bpm) in an otherwise uncomplicated pregnancy. Extensive ascites, a small pericardial effusion, umbilical venous pulsations, and ductus venosus flow reversal were noted. The cardiovascular profile score was 6/10 (Huhta et al, 2006). The heart size and biventricular function were normal and no structural cardiac defects were noted. fMCG proved invaluable in our diagnosis as, despite suspicion, we were unable to document AV dissociation on fetal echocardiograms. There was primarily 1:1 retrograde VA conduction with rare complete VA dissociation. Fetal intervals from flat averages during JET were: RP 82 ms; QRS 34 ms; QTc 493 ms. During sinus rhythm on follow up, they were: PR 116 ms; QRS 60 ms; QTc 456 ms. Treatment (digoxin/sotalol, then amiodarone) was successful in completely reversing the fetal ascites prior to delivery with normalization of ascites, UV pulsations, and DV flow reversal which had been present since presentation.
Conclusion: Treatment of suspected JET at any rate associated with ascites or abnormal cardiovascular profile scores is advisable. Where available, fMCG can prove to be an invaluable resource in the diagnosis, assessment of medication cardiac effects in the fetus, and characterization during successful treatment of a variety of cardiac rhythm abnormalities, especially if the diagnosis remains unclear with standard imaging modalities.

P1176 - NITROUS OXIDE GENERAL ANESTHESIA IN PEDIATRIC ELECTROPHYSIOLOGY PROCEDURES IS SAFE AND EFFECTIVE FOR REVEALING THE CLINICAL ARRHYTHMIA

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Background: During electrophysiology procedures, automatic atrial and ventricular arrhythmias, and sometimes AVNRT, can be suppressed by inhalation anesthetic agents such as sevoflurane. In pediatric patients, the option of moderate sedation is limited by patient acceptance and cooperation. In this study, we used nitrous oxide (N2O) in combination with versed and fentanyl (balanced technique) in cases where inhalation general anesthesia (GA) appeared to suppress clinical arrhythmia. N2O was chosen because of its sympathetic potentiation and absence of effect on cardiac conduction, and in most cases helped to reveal the clinical arrhythmia.
Methods: A retrospective review of 11 patients who underwent this GA technique was performed (May 2015 – July 2016). Patients were 11-19 years, with documented atrial tachycardia (AT), PVCs with high burden, AVNRT, or ventricular tachycardia (VT). In all cases anesthesia induction was with propofol, followed by conversion to balanced technique. Patients were monitored for anesthesia parameters and spontaneous occurrence of clinical arrhythmia, followed by standard EP testing and mapping/ablation.
Results: In 8 of 11 cases, N2O transition helped to reveal the clinical arrhythmia, which was initially suppressed (internal control). In six cases, the clinical arrhythmia was spontaneous after N2O transition, and inducible with stimulation protocols in the other cases. Arrhythmias included: AT (3), PVCs/VT (4), typical AVNRT (1). Ablation was successful in 6/8 cases, with arrhythmia mapping made possible by N2O transition in all cases. During the same time period, 6 patients underwent AT or VT/PVC ablation at our center where sevoflurane GA did not suppress the arrhythmia. The N2O technique provided stable hemodynamics and did not prolong emergence times. Post-op nausea and vomiting was seen in only 2 patients (16%).
Conclusions: In pediatric EP procedures where inhalational anesthetics suppress the arrhythmia, a balanced GA technique with N2O, fentanyl/versed is a safe and effective alternative to expose the clinical arrhythmia.

P1313 - CALMODULIN MUTATIONS CAUSE A SEVERE CLINICAL PHENOTYPE

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Background and Objectives: Cardiac channelopathies are inherited syndromes caused by mutations in genes encoding for ion channels, their subunits, or associated proteins that predispose the affected patient to arrhythmias, syncope and sudden death. Mutations in calmodulin, a ubiquitously expressed and essential calcium-signaling protein, have been recently associated with severe forms of Long QT Syndrome and Cathecolaminergic Polymorphic Ventricular Tachycardia, with life-threatening arrhythmias occurring at early age. Our aim here is to study the clinical behaviour of calmodulin mutations in a pediatric population with cardiac primary electrical disorders and compare it with channelopathies of a different genetic origin.
Methods: Children studied at our institution with a documented diagnosis of cardiac channelopathy that had been genetically tested were retrospectively included. For those without a previously identified genetic cause, CALM genes were sequenced. Clinical variables were collected and analyzed.
Results: 20 patients (mean age 11'5 ± 4'6 years) were included. 75% were male. Long QT Syndrome was the most prevalent diagnosis. 4 patients had an identified mutation in KCNQ1, 3 KCNH2, 2 SCN5A, 1 KCNJ2, 1 KCNQ1 + KCNH2, 3 in

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ryanodine receptor RYR2, 2 in calsequestrin CSQ2. 3 patients had a mutations in CALM: in 2 cases N98S in CALM2, in 1 case a novel missense mutation E141K in CALM3 was identified. No genetic mutation could be found in 1 patient. Patients with CALM mutations had an earlier onset of the disease (3'6 ± 3'5 vs 7'6 ± 3'7, p = 0,013) with a poorer prognosis in terms of cardiac arrest (all 3 cases) and sudden death (2 of 3) compared to channelopathies of a different genetic origin. All the mutations in CALM were the novo, with both parents unaffected.
Conclusions: Our results confirm previous studies suggesting that calmoduline mutations can cause an unusually severe phenotype with high risk of sudden death and very early age of onset.

Table 1.

	NO CALMODULIN MUTATION	CALMODULIN MUTATION
Numberofpatients:20	17 (85%)	3 (15%)
Male	14 (82%)	1 of 3
Ageofdebut	7,6 + /-3,7	3,6 + / 3,5
Symptoms (syncope, cardiacarrest ,death)	10 (58,8%)	3 of 3
Cardiacarrest	0	3 of 3
Death	0	2 of 3
ICDimplantation	5 (29%)	2 of 3
Numberofaffected familiars	2 + /-2,6	0
Deathinotherfamily members	9 (52%)	0

P1383 - REAL TIME THREE DIMENSIONAL ECHOCARDIOGRAPHY GUIDED ELECTROPHYSIOLOGICAL PROCEDURES IN PEDIATRIC PATIENTS

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Background/Hypothesis: Radiofrequency ablation of arrhythmogenic substrates is usually guided by three-dimensional (3D) electroanatomic mapping and fluoroscopy. Real-time 3D transesophageal echocardiography (RT3DTEE) has revolutionized interventional procedures by providing accurate visualization of various complex cardiac defects and intraprocedural guidance of catheters.
Materials and Methods: A 13-year-old female (Case 1) with Wolff-Parkinson-White syndrome and ventricle dysfunction due to right lateral accessory pathway; a 14 (Case 2) and 16-year-old male (Case 3) with incessant ectopic atrial tachycardia localized to the left atrial appendage (LAA); a 17-year-old male (Case 4) with atrial tachycardia arising from the aortic mitral continuity; and a 17-year-old male (Case 5) with ventricular tachycardia arising from the perimembranous septum underwent electrophysiological studies under RT3DTEE guidance. In another 17-year-old female presenting with intraatrial reentrant tachycardia status post Fontan procedure, RT3DTEE was used in conjunction with 3D electroanatomic mapping (Case 6).
Results: Visualization of the arrhythmogenic focus was enhanced by RT3DTEE and was useful in catheter positioning. The arrhythmogenic foci were successfully ablated in Case 1 and 3-6. Although the focus of the arrhythmia was accurately identified in Case 2, ablation was not successful due to its epicardial location. Real-time monitoring of cryoablation by demonstration of ice balling and evaluation of dyssynchrony was made possible by RT3DTEE in Case 1. Additional anatomic details like juxtaposition of atrial appendages were also easily observed in Case 1.
Conclusions: Arrhythmogenic substrates can be accurately identified on RT3DTEE, which provides enhanced visualization and serves to prevent complications by reducing radiation exposure. When combined with 3D electroanatomic mapping, RT3DTEE augments visualization of arrhythmogenic substrates and has a role for electrophysiological procedures in the setting of complex CHD.

P1460 - CLINICAL AND GENETIC PROFILE OF CONGENITAL LONG QT SYNDROME IN HONG KONG – 18 YEAR EXPERIENCE

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Background: Comprehensive review of all patients diagnosed with congenital long QT syndrome (LQTS) in a single tertiary paediatric cardiology centre is described.
Methods: All paediatric patients diagnosed at our centre with LQTS from January 1998 to December 2016 were included. LQTS was established with either a Schwartz score >= 3.5 or presence of pathogenic mutation.
Results: 59 subjects (33 boys) fulfilled the study inclusion criteria, with a mean age of 8.90 + /- 5.74 years at diagnosis. The mean follow-up duration was 5.33 + /- 4.65 years. 84.7% individuals were probands. 23 (39.0%) and 7 (11.9%) subjects presented with syncope and convulsion respectively. 7 subjects (11.9%) had cardiac arrest on presentation. Fetal bradycardia and neonatal 2:1 atrioventricular block were also the mode of presentation in 2 subjects. The mean corrected QT interval in our cohort was 504 + /- 47 ms. 42.4% individuals had a positive family history. 37 subjects (62.7%) confirmed to have pathogenic mutation for LQTS (LQT1-16.9%; LQT2-18.6%; LQT3-11.9%; LQT5-5.1%; LQT8-10.2%). No deaths were reported but 2 subjects had subsequent cardiac arrest despite on metoprolol. Pacemaker was implanted in 4 subjects, while implantable cardioverter-defibrillator was implanted in 14 subjects. 2 patients underwent left cardiac sympathectomy. 64.4% individuals were put on medical therapy, with metoprolol being the initial choice of beta-blocker therapy in half of our cohort.
Conclusions: 18-year experience in management of paediatric LQTS in a tertiary cardiology centre is described, with a significant proportion of probands. Syncope and convulsion were the main forms of presentation. Use of metoprolol was associated with breakthrough attacks in children with LQTS. Our cohort demonstarted relative good survival.

P1607 - CLINICAL COURSE AND ELECTROPHYSIOLOGICAL CHARACTERISTICS OF BINODAL DISEASE AT CHILDREN WITH IMPLANTED PACEMAKERS

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Purpose: This study is aimed to assess the results of the clinical course and electrophysiological characteristics of binodal disease (BB) at children with implanted pacemakers.
Methods: We enrolled 40 consecutive patients with BB who underwent pacemaker implantation, 24 (60%) male, 16 (40%) female. The average age for the moment of pacemaker implantation - 13,84 ± 0,621 (2,8-17,9) years. The longevity of persistence of BB before the pacemaker implantation had been 5,31 ± 4,36 years (several days - 14,29). BB included: combined sinus node dysfunction (SND): sinus bradycardia, sinoatrial block, sinus arrest with escape ectopic rhythm, and/or increasing the sinus node recovery time (SNRT) and/or corrected SNRT (cSNRT) and atrioventricular (AV) conduction disturbance as AV block of grade I or greater, and/or Wenckebach point (WP) below 500 msec. All children had carried out examination of the heart at least 2 times: ECG, Holter monitoring (HM), transesophageal electrophysiologic study (TEEPS) before pacemaker implantation and ECG, HM, noninvasive electrophysiologic study after pacemaker implantation.
Results: Long-term follow up of patients was 52,2 + 35,22 months (range 2,7 to 145,9). There was a statistically significant increase in the duration of SNRT 1617,7 ± 441,6 ms at the first examination to 1915,37 ± 655,7 ms at the end of follow-up ($p = 0.04$). In addition, there was a significant decrease in the Wenckebach point of 117 ± 35.52 pulses/min. to 102,2 ± 30,42 pulses/min. ($p < 0,05$).
Conclusions: Among children with binodal disease who underwent pacemaker implantation in monitoring there has been showed an improvement of clinical and hemodynamic picture of the disease during the follow-up, but in the whole the group experienced a worsening of electrophysiological parameters.

P1608 - ARRHYTMIAS AND CONDUCTION DISORDERS IN PATIENTS WITH SURGICAL CORRECTION OF ANOMALOUS TOTAL PULMONARY VENOUS CONNECTION AT THE NATIONAL INSTITUTE OF CARDIOLOGY "IGNACIO CHAVEZ"(2005 214)

Emilia Patino-Bahena[1], Arianna Covarrubias[1], Pablo Mendoza[2], Antonio Benita-Bordes[3], Samuel Ramírez-Marroquin[3], Alfonso Buendía-Hernandez[1]
National Institute of Cardiology, Paediatric-Cardiology, México-Mexico[1]; National Institute of Cardiology, Arrhythmias, México-Mexico[2]; National Institute of Cardiology, Surgery, México-Mexico[3]

Congenital heart defects are associated with arrhythmias, not only for the congenital anatomic component, there is also an increasingly acquired arrhythmogenic substrate associated with surgical repair. For the formation of new tachycardia circuits in the atriotomy, patches or prostheses and suture lines in approximately 10- 20 years of post-surgical evolution. A retrospective, descriptive, observational study of patients with anomalous total pulmonary venous connection who underwent total correction and subsequent arrhythmias after surgery at the National Institute of Cardiology "Ignacio Chavez" from January 2005 to December of 2014. We found 223 patients with this diagnosis after total correction. In 107 patients (48%) some type of rhythm or conduction disorder was documented. Conduction abnormalities were the most frequently found alteration, being nodal rhythm and complete atrioventricular block the predominant presentations in the short and medium term; and in the late period the most frequent abnormality was sinus node dysfunction. Ventricular fibrillation was the main tachyarrhythmia in the short and medium term, it should be mentioned that it is a poor prognosis that is associated with death; in the late period the atrial flutter. The main therapeutic intervention was the temporary pacemaker implantation. Eight patients required definitive pacemaker. Cardiac ablations were performed in 2 patients for atrial flutter.

P1631 - CLINICAL UTILITY WITH AN OBSERVATION OF ATRIAL KICK WAVE PATTERNS FOR DIFFERENTIAL DIAGNOSIS OF FETAL EXTRASYSTOLE

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Generally, obstetricians tend to have a feeling of being not good at fetal arrhythmia on ultrasound. If fetal electrocardiography becomes clinical widespread, we will be able to diagnose fetal arrhythmia accurately. However, at present we have to do diagnosis of fetal arrhythmia using echocardiography, M-mode method and superior vena cava/aorta Doppler flow method (SVC/AA Doppler method), because of no clinical widespread of fetal electrocardiography. This time, we hypothesize that the characteristic Doppler flow pattern exists in each extrasystole, premature atrial contraction (PAC) and premature ventricular contraction (PVC). We investigate how to better diagnose each extrasystole by those Doppler flow patterns. In this study, we enrolled 26 children (14 males and 12 females between 0 and 17 years old), who were already diagnosed with PVC or PAC using 12-lead ECG and Holter ECG. We grouped into 2 (19PVCs and 7PACs) and analyzed Doppler flow patterns using modified SVC/AA Doppler method. As the result, we find the following Doppler flow patterns; 1) in PVCs group, mostly similar reversal flow exists at midpoint of reversal flows (atrial kick wave) before and after extrasystole, 2) in PACs group, no exists at midpoint. In conclusion, observation of atrial kick wave patterns using SVC/AA Doppler method might be clinically useful for differential diagnosis of each extrasystole in fetus.

P1652-3 DIFFERENT ICD INDICATIONS IN CHILDHOOD

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The ICD is the first-line treatment and prophylactic therapy for patients at risk for sudden cardiac death due to ventricular fibrillation and ventricular tachycardia. Recommendations for secondary prevention in children are similar to ICD implantation guidelines developed for adults, despite a paucity of pediatric randomized controlled trials. In this poster we reported 3 different ICD indications in childhood, which were implanted in our institute. CASE 1A9-year-old male patient who had HCMP diagnosis was admitted to the pediatric intensive care unit after resuscitation because of cardiac arrest. The emergency team stated that while the patient found there was cardiac arrest and the monitorization showed asystole. In intensive care unit no rhythm abnormality was detected except septal hypertrophy in ECG (image 1). Because of presented asystole and suspected VT an ICD implanted and discharged after 5 days of follow-up. CASE 2 A 14-years old male patient admitted hospital because of seizures. In pediatric intensive care unit, ventricular tachycardia was detected at the time of seizures. While receiving IV inotropes frequent VT episodes were detected in 24 hour holter rhythm monitorization. And this condition is diagnosed as Catecholaminergic polymorphic ventricular tachycardia (image 2). IV inotropes were

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stopped, propranolol initialized. After rehabilitation and decreasing the VT episodes ICD implanted, and discharged with oral propranolol. CASE 3 A 10-years old female patient admitted hospital because of recurrent syncope attacks. ECG revealed sinus rhythm, but QTc interval was calculated 0.50 msn (image 3). Within 24 hours ECG holter monitorization, sustained VT attacks were detected. So that we gave propranolol therapy and implanted ICD.

Discussion: Patients who have survived after sudden cardiac arrest due to malignant arrhythmias would benefit from ICD implantation. The indications for ICD implantation in pediatric patients and those with congenital heart disease have developed in recent years with adult randomized clinical trials. Similar to adults, ICD indications have evolved from secondary prevention of cardiac death.

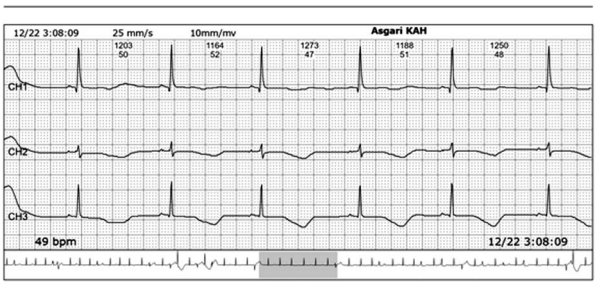

Figure 1.

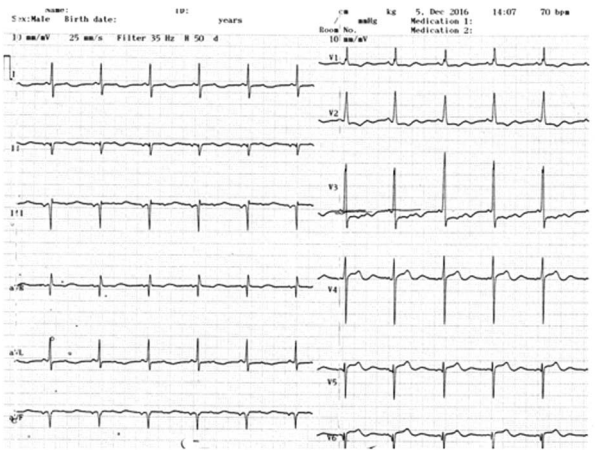

Figure 2.

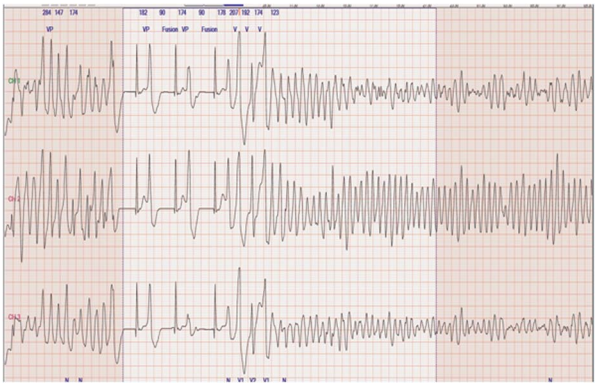

Figure 3.

P1686 - PERMANENT PACEMAKER IMPLANTATION VIA ILIAC VEIN IN SINGLE VENTRICLE PATIENT AFTER BIDIRECTIONAL GLENN PROCEDURE

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University of Texas Health Science Center at Houston/Children's Memorial Hermann Hospital, Pediatric CV Surgery, Houston-United States[2]

Background: We report transvenous permanent pacemaker placement (PPM) via Iliac vein after failure of epicardial pacemaker system in a patient (pt) with single ventricle (SV) post bidirectional Glenn (BDG) procedure.

Methods: N/A

Results: A 23-year-old morbidly obese female with DS, tricuspid atresia, post BDG procedure as destination palliation presented with one-week history of marked bradycardia with ventricular escape rate between 20-40 bpm. Prior attempt at epicardial PPM placement for history of advanced atrioventricular block was complicated by high pacing thresholds and pocket infection. Attempt to replace pulse generator (PG) which had reached end-of-life and revise non-functioning epicardial ventricular pace-sense lead was unsuccessful and aborted following significant bleeding-related complications. Alternative approaches to transvenous PPM were discussed. An incision was made superior to inguinal ligament and right external iliac vein was cannulated with two long introducer sheaths. Two custom size pace-sense leads (CapSureFix Novus MRI, 85 cm, Medtronic Inc.) were advanced through each of these sheaths and actively fixated into lateral wall of right atrium and left ventricular apical septum achieving optimal pacing thresholds, impedance and sensing margins. Figure demonstrates lead positioning. An abdominal para-umbilical subcutaneous pocket was created for PG and leads were tunneled from the inguinal incision into this pocket. There were no procedural complications. Combination of Aspirin and Clopidogrel was started for thrombotic prophylaxis. Pt developed non-oliguric renal dysfunction related to prophylactic antibiotics following procedure, which did not require any intervention. She had been doing well at her two-months follow-up with normal pacemaker function.

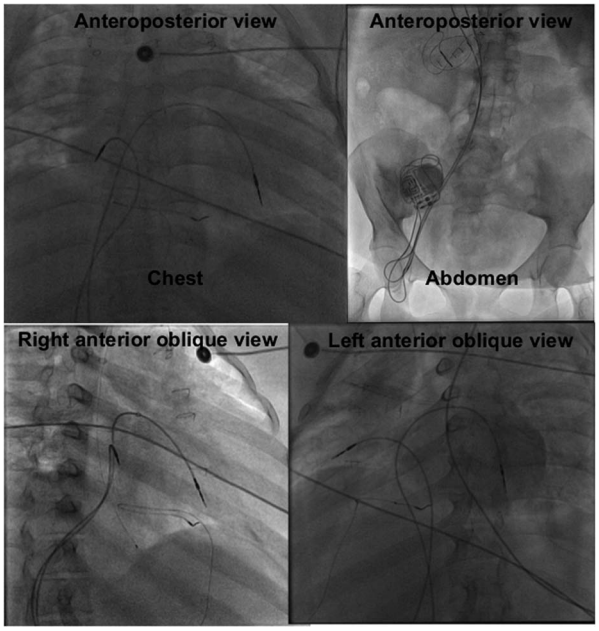

Figure 1.
Fluoroscopic images.

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Conclusions: PPM with trans-iliac vein leads tunneled to abdominal PG offers a less invasive and feasible alternative to epicardial pacemaker system in palliated SV pts, when direct access via upper venous tree is not available post BDG procedure. This approach offers the benefit of unrestricted pt ambulation, optimal pacing thresholds with lowered risk of thromboembolism on anti-platelet agents.

P1691 - EARLY POSTOPERATIVE SUPRAVENTRICULAR TACHYCARDIA AFTER CONGENITAL HEART DISEASE SURGERY

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Background: Postoperative supraventricular tachycardia (SVT) after congenital heart disease (CHD) surgery was occasionally seen in early postoperative period but its clinical significance was not clear.
Methods: From 2010 to 2015, all consecutive patients, aged less than 18 years-old, who had early postoperative SVT after cardiopulmonary bypass surgery for CHD in our institution were reviewed. The definition of SVT includes atrial arrhythmia and paroxysmal supraventricular tachycardia.
Results: Totally 29 patients (M/F 19/10) were enrolled. Median age of operation was 4 months (range 1 day – 17.55 years), with 62% of the patients less than 1 year-old. The common underlying cardiac anomalies include complex CHD in 12, transposition of great arteries in 5, and tetralogy of Fallot in 3. Among them, 8 (27%) patients had history of previous cardiac surgery, and 5 (17%) patients had SVT history. Most of the patients with SVT history had multiple cardiac surgery before (Odds ratio 2.76, $p = 0.027$). The pre-operative condition was hemodynamically unstable in 13 patients (45%) with 2 required extracorporeal membranous oxygenator support after surgery. The mean atrial and ventricular rates were 278 ± 89.8 bpm and 196 ± 41.2 bpm, respectively. The SVT was controlled by single dose IV adenosine in 5, IV amiodarone in 17. Only 2 patients need DC cardioversion due to critical hemodynamics. Compared to those requiring IV anti-arrhythmia agents more than 24hr, those who use less than 24hr had shorter ICU stay (14.3 vs. 80.8 days, $p = 0.0073$). Maintenance oral antiarrhythmic agents were needed in 27 patients: 11 required multiple antiarrhythmic agents. The median duration of oral antiarrhythmic agents was 167 days (7-2072 days), and the medication could be discontinued in 18 patients with only one recurrence.
Conclusions: Early postoperative SVT is not uncommon after CHD surgery especially for those with complex CHD and cyanotic CHD. It may cause hemodynamically compromise but can be controlled by medication.

P1700 - ELECTRICAL ABNORMALITIES IN CHILDREN WITH LAMINOPATHIES

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Children's Hospital,Juan P Garrahan, Cardiology Department, Buenos Aires-Argentina[1]; Children's Hospital,Juan P Garrahan, Cardiac Catheter Laboratory, Buenos Aires-Argentina[2]; Children's Hospital,Juan P Garrahan, Neurology Department, Buenos Aires-Argentina[3]

Background: Lamin proteins are type V intermediate filaments, a mayor component of the nuclear lamina that supports the inner nuclear membrane. Laminopathies (LMNA) mutations have been reported associated to distinct phenotypes, collectively termed laminopathies. These, usually affect skeletal and/or cardiac muscle, white fat distribution, bone and peripheral nervous tissue, and also may cause premature aging with or without myopathy.
*Objectives:*To describe the electrical abnormalities,treatment and mortality in patients with early onset of LMNA-related muscular dystrophies.
Methods: Between 2008-2015, sixteen patients(p)underwent a cardiac check-up with electrocardiogram (EKG), 24 hour Holter monitoring,and transthoracic colour Doppler echocardiography. Median follow- up was 4,3 years(IQR 1,25-6,45).
Results: The mean age onset of symptoms was 2.13 + /-1.7 5 years, and the initial electrical disturbance appeared at10.22 + /-2.39 years. EKG findings: P wave amplitude less than 0.1 mV in 8(53,3%), AV block: in 4(26,6%), bundle branch block in 4, premature ventricular contractions in one and abnormal repolarization in 2 patients. Holter monitoring reported: premature atrial contractions 20%(3p), atrial tachycardia 26,6%(4p). Ventricular arrhythmias included simple premature ventricular contractions 20%(3p), with a pattern of bigeminy,couples and non- sustained ventricular tachycardia. Sick sinus syndrome 6,6% (1p); and second degree AV block Mobitz type I 13,2%(2p)were also documented. Antiarrhythmic treatment: Two patients received amiodarone, betablockers 2 and a combined treatment 2p. A pacemaker was implanted in three patients at 16 and 17years. The indications were sick sinus syndrome and progressive AV block. Three patients died during follow-up. In the univariate analysis, mortality was related to history of sustained atrial arrhythmias (p 0.0001), progression of AV block (p0.0001) and ventricular tachycardia (p 0.01).
Conclusions: The most common electrical disturbances in patients with laminopathies were: progressive decrease in P-wave amplitude, first degree AV block and second degree AV block Mobitz I type and atrial and ventricular arrhythmias. The electrical disturbances developed later than the neuromuscular symptoms. Mortality was related to second degree AV block, sustained atrial tachycardia with decreased left ventricular function and ventricular tachycardia.

P1737 - SINUS ARREST AS UNREPORTED PRESENTATION OF KCNE1 MUTATION

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Queen Elizabeth Hospital, Paediatrics, Hong Kong-Hong Kong[1]; Queen Mary Hospital, Paediatric Cardiology, Hong Kong-Hong Kong[2]; Queen Mary Hospital, Paediatric and Adolescent Medicine, Hong Kong-Hong Kong[3]

We report a 6-year-old boy who presented with recurrent episodes of unprovoked syncope. His 12-lead electrocardiogram (ECG) demonstrated borderline prolonged QT intervals, with maternal ECG also showed similar findings. Instead of ventricular arrhythmia, sinus arrest with pauses up to 7 seconds was identified as the cause of recurrent syncope, with the use of implantable loop recorder (ILR). Genetic testing revealed a pathogenic mutation in KCNE1, which is known to cause long QT syndrome type 5. Implantable cardioverter-defibrillator was implanted. The early use of ILR allows early detection of underlying arrhythmia and allows timely intervention. This unreported link between sinus arrest and KCNE1 mutation is also discussed.

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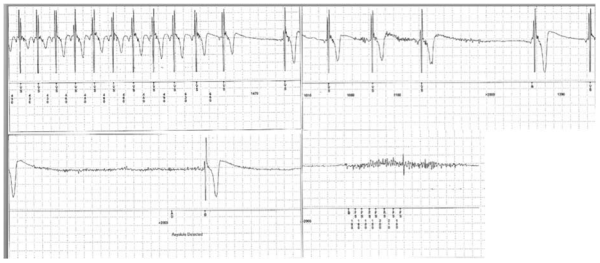

Figure 1.

P1772 - USEFULNESS OF STRESS TESTING IN THE DIAGNOSIS OF CATECHOLAMINERGIC POLYMORPHIC VENTRICULAR TACHYCARDIA

Juan Villafane
University of Kentucky, Pediatrics (Cardiology), Louisville-United States

Background: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a cardiac channelopathy, which may manifest with sudden death early in childhood. Symptoms are associated with emotional stress or exertion.
Case report: A 4.5-year-old female with history of two abrupt syncopal spells during moderate exertion – one of them resulting in head trauma and unresponsiveness for 30 seconds. No detectable seizure activity. EMS was called and CR monitoring did not show any arrhythmias. The patient was referred to a neurologist who insisted on getting a cardiovascular evaluation. Resting ECG was normal. Echocardiogram showed a small interatrial communication. Holter showed normal heart rates (HR). Ventricular bigeminy and multiform couplets were detected exclusively when HR was over 130 bpm. Stress testing was performed based on the history of exertional syncope and ventricular ectopy. Bidirectional premature ventricular contractions (PVCs) appeared at HRs above 122 bpm. Ventricular ectopy became progressively worse, including ventricular bigeminy followed by non-sustained CPVT. There was no ventricular ectopy when HR dropped <120 bpm. At the end of the test, there was sinus rhythm with normal AV conduction, QT intervals, and T wave morphology. The patient was started on nadolol. Follow-up Holter was unremarkable except for 11 isolated PVCs at HR >120 bpm. There was a brief episode of ventricular bigeminy at HR > 137.
Conclusions: CPVT is a malignant channelopathy that can present in the very young. A child with exertional syncope, a normal QT interval and exercise-induced ventricular ectopy is more likely to have CPVT (than long QT syndrome). Stress testing should be attempted at any age in this scenario.

P1790 - SUPRAVENTRICULAR MICRO REENTRY IN A NEWBORN DUE TO A GIANT ATRIAL SEPTUM ANEURYSM

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Background/Hypothesis: The focal micro reentry tachycardia (FMRT) is a rare form of atrial tachycardia in children and belongs to the group of supraventricular tachycardias (SVT´s). The reentry circuit is mediated via areas of functional conduction block, occurring in adjacent tissues with differing refractory periods, and the symptoms range from an asymptomatic patient to severe heart failure. The treatment depends on the mechanism of the arrhythmia and the clinical state of the patient. This case is the first one of a newborn presenting with a FMRT possibly associated to a giant atrial septum aneurysm (ASA).
Case Presentation: A 13-days-old male patient was referred to our center due to recurrent tachycardia (240 bpm) in a stable clinical condition. In our center supraventricular reentrant tachycardia was diagnosed after application of adenosine (0,2 mg/kg). Echocardiographically an atrial septal defect with a giant ASA and a very small muscular ventricular septum defect were diagnosed; cardiac function was normal. The SVT could successfully be terminated by the use of electric cardioversion with 5 Joule (2 Joule/kg). A relapse after 3 hours could again be terminated with an electric cardioversion (6 Joule). Therefore we started the substitution of digoxin and sotalol (112 mg/m^2). With this therapy the patient could be discharged in good clinical condition after 4 days. After six months without the history of relapses, the medication has been discontinued. Due to the elucidated pathomechanism described above, we assume a correlation between the ASA and the FMRT.
Conclusion: This is the first report of a newborn with a focal atrial micro reentrant tachycardia, assumingly related to a giant atrial septum aneurysm. The tachycardia was effectively treated by cardioversion and medicinal prophylaxis with digoxin and sotalol.

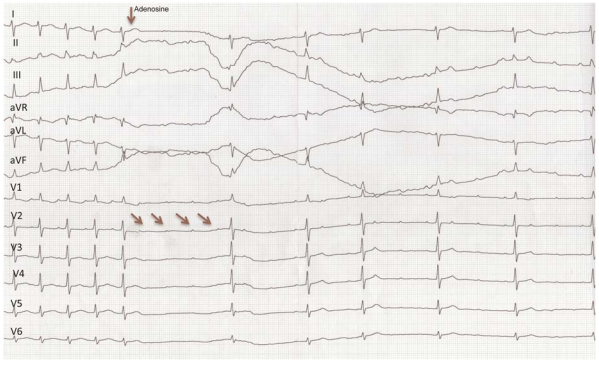

Figure 1.

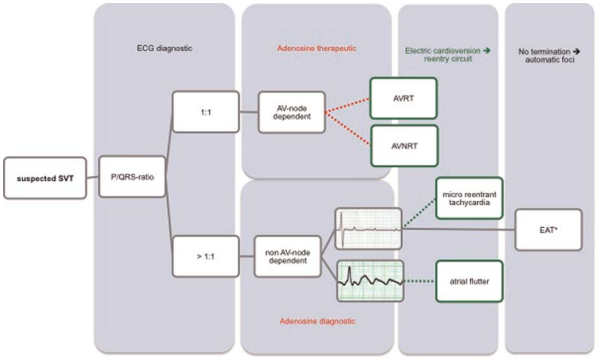

Figure 2.

P1801 - PROJECT ADAM PREVENTION OF SUDDEN CARDIAC DEATH IN CHILDREN AND ADOLESCENTS THE SUCCESSES OF A NATIONAL PROJECT OF SECONDARY PREVENTION INCLUDING CPR AED EDUCATION BROAD NATIONAL COVERAGE AND COLLABORATION TO SAVE LIVES

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Background: Sudden cardiac death (SCD) in children and adolescents is devastating. Many SCD events occur in schools, which represent community gathering places. A comprehensive program of CPR-AED education of students and staff, emergency preparedness, and AED placement in schools could improve the low bystander CPR rate (20-30%) and out-of-hospital cardiac arrest (OHCA) survival rate (10%).
Materials and Methods: A statewide three-tiered approach to secondary prevention was instituted in Wisconsin in November 1999 with the goals of: (1) Providing education regarding SCD presentation and layperson response, (2) Advocating for mandatory CPR-AED education for students prior to graduation, and (3) Placing AEDs in all schools and playing fields/arenas.
Results: Since the original Project ADAM program in Wisconsin began, 12 other states have joined Project ADAM as affiliate programs. Key elements for a successful program have included administration ownership by children's hospital or academic medical center with a pediatric cardiac program, a physician medical director, and a program director/coordinator. In addition, Project ADAM provides a template that accommodates the variability in affiliate programs with regards to resources, infrastructure, funding, and state laws. Affiliate programs recruit local champions to advocate for the development of emergency action plans, school CPR-AED programs, education about effective CPR and AED use, and funding and maintenance of AEDs. Project ADAM has resulted in greater than 120 lives saved, both children and adults. Additionally, Project ADAM has provided advocacy resulting in the passage of state legislation mandating CPR and AED instruction as a high school graduation requirement.
Conclusions: Project ADAM is a school-based secondary prevention program for SCD that has saved lives of children and adults. Adopted nationwide, such an approach has the potential to significantly improve the relatively poor outcomes associated with OHCA and will likely result in a greater incidence of high quality bystander CPR.

P1892 - NEONATAL ARRHYTHMIAS AND INDEXES OF HEART RATE VARIABILITY IN NEWBORNS

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Aim: to compare the frequency of occurrence and variants of cardiac arrhythmia in full-term and preterm newborns.
Methods: analysis the data of anamnesis, Holter monitoring ECG (Holter ECG), Doppler echocardiography, statistical analysis.
Results: In 79 infants had irregular heart rhythm. The 1st group was included prematurely born children 55 (69.6%), the 2nd group - full-term newborns (24 - of 30.4%). Hypoxic-ischemic CNS damage moderate, and severe degrees of gravity was found in 29 (52.7%) of the 1st group and in four children (29.2%) the 2nd group ($t = 3.13$, $p < 0.05$). Central hemodynamics according to Doppler echocardiography showed the presence of hyperkinetic and hypokinetic types of hemodynamics in almost equal shares (45.8% and 41.7%, respectively) in a group of full-term infants. Significant differences between the groups was observed in patients with normokinetic type of hemodynamics ($t = 5.5$, $p < 0.05$). According to the results of Holter ECG among cardiac arrhythmias have been identified in both groups: atrial extrasystoles – 40.5%, ventricular extrasystoles – 13.9%, episodes of transient AV-block of 1st degree – 10.1%, long QT – 10.1%. No significant differences were between groups in the frequency of occurrence of AV-block of 2nd degree type Mobitz 1 in 4 preterm babies ($7.3 \pm 2.2\%$, $t = 3.13$, $p < 0.05$). Processing of the obtained statistical indices of heart rate variability non-parametric methods have revealed a reliable decrease in SDNN in the group of premature born children ($p < 0.05$). SDNN displays the decrease of the total effect of autonomous regulation of circulation that is associated with the strengthening the regulation of cute, depressing the activity of the autonomous circuit.
Conclusions: A rhythm disorder of the heart in preterm babies was connected to the influence of hypoxic lesions of the CNS in the regulation of cardiac activity largely and to the complexity of the flow adaptation processes.

P1912 - LONG TERM RESULTS OF RADIOFREQUENCY CATHETER ABLATION IN CHILDREN WITH ATRIOVENTRICULAR NODAL REENTRANT TACHYCARDIA

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Background: There are no reports regarding atrioventricular nodal reentrant tachycardia (AVNRT) after RFCA in children. Therefore, we evaluated long-term results of AVNRT after RFCA in children.
Material and Methods: From 1993 to 2001, 71 children underwent RFCA of AVNRT at our institution. We conducted a questionnaire survey for 52 patients and obtained replies from 23. The contents of the questionnaire were follows; Have you ever experienced palpitation after catheter ablation? Did you visit a hospital after treatment because of the recurrence of palpitation or arrhythmia? Are you satisfied with the efficancy of this treatment? Based on the replies, we analyzed the patients' age, sex, body mass index, history of medications, congenital heart disease, one echo after RFCA, and types of AVNRT. Twenty-three patients were divided in two groups; Group A comprised 12 patients with the present of palpitations and Group B comprised 11 patients with absence of palpitations after RFCA.
Results: All patients had experienced palpitations before RFCA; 12 (52%) currently experienced palpitations after RFCA; seven (30%) visited a hospital after RFCA; and 22 (96%) were satisfied with the efficancy of the treatment. The number of patients visiting a hospital after RFCA was significantly higher in Group A (58%) than in Group B(0%), and the frequency of satisfaction by this treatment showed no apparent differences (Group A, 92%; Group B, 100%). In Group A, two patients experienced recurrences. there were significant differences in the recurrence of RFCA between the two groups (A: 58%, B: 17%) with regard to history of medications, whereas others had no differences.
Conclusions: More than half of patients with AVNRT experienced palpitations without recurrences after RFCA. Most of the patients were satisfied with the treatment. In this study, the exact cause of palpitations was unknown; however, palpitations after RFCA might be involved in history of medications.

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P1980 - AMBULATORY ARRHYTHMIA DETECTION WITH ZIO® XT PATCH IN PEDIATRIC PATIENTS

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Introduction: A novel device, ZIO® XT Patch (ZIO), can be utilized for ECG monitoring for 14 days. The purpose of this study is to describe the duration of ZIO use by age, as well as time to arrhythmia detection in the pediatric population.
Methods: A single-center, eview of patients ≤17 yrs prescribed a ZIO from 10/14 to 2/16. All data were analyzed, along with duration of ZIO use and time to first arrhythmia. Arrhythmia was defined as supraventricular tachycardia (SVT), advanced atrioventricular block (AV block), or ventricular tachycardia (VT). All ZIO reports were reviewed by a blinded pediatric electrophysiologist. Comparisons were made to 24-hour Holter monitors from the same time period.
Results: A total of 406 ZIO monitors were prescribed for 363 patients; median age 12.7 yrs (0.01-17 yrs), 50% male. The median duration of monitoring significantly increased with age: 1 day (1-14) for 41 ZIO in age <3 yrs, 2 days (1-13) for 57 ZIO in age 3-6 yrs, 3 days (1-14) for 109 ZIO in age 7-12 yrs, and 4 days (1-14) for 199 ZIO in age 13-17 yrs ($p < 0.001$). A total of 499 Holter monitors were performed; median age 4.9 yrs (0.03-17 yrs), 51% male. Median age differed between the ZIO and Holter groups ($p < 0.0001$), but rates of detection were similar: 11% by ZIO vs 10% by Holter ($p = 0.51$). Arrhythmia was identified on: 45 ZIO (35 SVT, 3 AV block, 7 VT) and 48 Holter (34 SVT, 3 AV block, 11 VT). The first arrhythmia on ZIO occurred in <24 hrs of monitoring in 42% patients, 24-71 hrs in 24%, and >72 hrs in 33%, while arrhythmias detected by Holter.

P1981 - CORRELATION BETWEEN ECG ABNORMALITIES AND MARIJUANA USE IN THE PEDIATRIC POPULATION

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Purpose: The effects of marijuana on the cardiac conduction system are ill defined. The purpose of this study is to describe the association between electrocardiogram (ECG) findings and positive drug screening (+ UDS) for marijuana in the pediatric population.
Methods: A retrospective review was conducted from charts dated 10/13-11/14 of patients ≤18 years of age that tested positive for marijuana exposure by urine screen in the Emergency Department (ED). All ECGs performed were reviewed by two blinded pediatric cardiologists.
Results: There were 174 patients identified in the ED with a + UDS with a median age of 15 yrs (0-18 yrs); 42% were male. ECG at time of + UDS was performed on 37 (21%) patients. Abnormal ECG finding was identified in 16/37, of which 15 had another ECG performed on a different date. Comparisons were made between these ECGs; significant differences were noted in those patients with + UDS, including ST segment changes (4 patients), left ventricular hypertrophy (3), and one each: atrial fibrillation, QT prolongation, Mobitz type I block, and right bundle branch block.
Conclusion: Abnormal ECG findings, including serious rhythm disturbances and conduction abnormalities can be identified in pediatric patients under the influence of marijuana. An ECG should be considered on all patients with a positive urine drug screen for marijuana.

P1995 - NOVEL TECHNIQUE OF ATRIAL MAPPING THROUGH PULMONARY ARTERY IN PATIENTS PALLIATED FOR SINGLE VENTRICLE CIRCULATION

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We present three cases where a catheter placed in the pulmonary artery (PA) provided clear atrial signals to assist in performing electrophysiological interventions on children palliated for single ventricle circulation.
Case 1: 7 year old girl with double outlet right ventricle, malposed great arteries and hypoplastic left ventricle had undergone cavopulmonary shunt, reduction atrioplasty of right atrium, ablation of incessant focal atrial tachycardia, tricuspid valve replacement and maze procedure. She underwent further electrophysiological studies(EPS) for recurrent flutter. A deflectable quadripolar catheter placed in left PA through internal jugular vein access gave clear atrial signals.
Case 2: 4 year boy with hypoplastic left heart syndrome(HLHS) underwent total cavopulmonary connection with extracardiac fenestrated conduit. He developed incessant atrial tachycardia in the post-operative period unresponsive to medical therapy. A quadripolar catheter placed in right PA gave excellent atrial signals.
Case 3: 3 year old boy with HLHS underwent Norwood procedure and superior cavopulmonary anastomosis had significant right ventricular dysfunction and was noticed to have 2:1 conduction block with 9 second pause for which he underwent epicardial dual chamber pacemaker. He developed atrial tachycardia for which he underwent EPS. A catheter placed in right PA gave excellent atrial signals and could also be used for atrial pacing. Atrial signals from the pulmonary artery were used as reference and for activation mapping using CARTO mapping system in all three patients and for pacing in two patients. There was acute success in two patients whilst the third patient had recurrence despite successful ablations.
Conclusion: Placement of the diagnostic electrophysiology catheter in the PAs is a useful technique for EPS involving children who have undergone cavopulmonary shunt or Fontan completion. This technique can be used to get atrial signals without entering the atrial mass and alternative venous access can be used if there is femoral vein occlusion.

P2031 - TRANS CATHETER BASED ELECTROPHYSIOLOGICAL MANAGEMENT OF ARRHYTHMIAS IN ACHD SINGLE CENTRE EXPERIENCE

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Introduction: Catheter ablation is increasingly used main modality for management of arrhythmias in adult congenital heart diseased (ACHD) population. Literature evidence supporting use of such treatment is based on case-reports and centre experiences. We report three-year data for consecutive ACHD patients undergoing ablation procedures in our centre.
Methods: Retrospective analysis of consecutive arrhythmia ablation procedures in ACHD patients between January 2014 and December 2016. Acute outcome was defined as: (1) Successful: termination of sustained clinical tachycardia by ablation and non-

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inducibility afterwards; or achieving pulmonary vein isolation (PVI)or cavotricuspid isthmus (CTI) ablation in sinus/paced rhythm; (2) Equivocal: intra-procedure degeneration of clinical arrhythmia into atrial fibrillation (AF) requiring cardioversion followed by substrate modification in sinus/paced rhythm (3) Failed: inability to terminate clinical tachycardia by ablation (excluding PVI); ending the procedure with cardioversion (excluding PVI); or if clinical arrhythmia remained inducible.
Results: 52 ablation procedures were performed in 43 patients (mean age: 41.3y). Underlying ACHD/repair was ASD/VSD: 12 patients (23%), Ebstein: 5(10%), Tetralogy of Fallot/pulmonary stenosis repair: 9(17%), Fontan repair: 9(17%), arterial switch: 3 (6%), Senning: 3(6%), Rastelli: 1(2%), Eisenmenger: 1(2%) and combinations: 9(17%). Average number of arrhythmias/patient was 1.2 (range: 0-4). Ablation was performed for CTI flutter in 20 patients (38%), AVNRT in 3(6%), AVRT in 3(6%), AF in 7(13%), intra-atrial re-entrant tachycardia (IART) in 8(15%), focal atrial tachycardia in 1(2%) and RVOT ectopics in 2(4%). Substrate modification was performed in sinus/paced rhythm in 13 patients (25%). Ablation outcome was successful in 45 cases (86.6%), equivocal in 2(3.8%) and failed in 5(9.6%). There was 1(2%) complication - asystolic arrest following termination of IART by ablation- successfully resuscitated. Average procedure time was 205 min (median 195). Mean hospital stay was 3.6 nights (median: 1).
Conclusion: Our data support that catheter ablation in ACHD patients is safe and has high acute success rate.

P2032 - DYNAMIC QT INTERVAL CHANGES FROM SUPINE TO STANDING IN HEALTHY CHILDREN

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Chu Sainte Justine, Pediatric Cardiology, Montreal-Canada[1]; Boston Children's Hospital, Electrophysiology, Boston-United States[2]; Montreal Heart Institute, Electrophysiology, Montreal-Canada[3]

Background: QT interval variations in response to exercise-induced increases in heart rate (HR) have been reported in children and adults in the diagnosis of long QT syndrome. A quick standing challenge has been proposed as an alternative provocative test in adults, with no pediatric data yet available.
Objective: Characterize physiological QT interval variations during a quick standing test in a cohort of healthy children.
Methods: A standing test was performed in 47 healthy children (mean age 9.3 ± 2.9 years) following 10 minutes in a supine position with continuous ECG recording. QT intervals were measured at baseline, maximal HR, maximal QT, and at each minute of a 5-minute recovery. Measurements were taken in leads II/V5 and corrected for HR (QTc).
Results: Upon standing, HR increased by 26 ± 9 bpm. QT interval was similar at baseline and upon standing (388 ± 32 versus 383 ± 34 msec, $p = 1.0$). However, QTc increased from 429 ± 21 to 493 ± 36 msec ($p < 0.001$). The 95th percentile for QTc at baseline and maximal HR was 457 msec and 550 msec, respectively. At 1 minute of recovery, QT interval was shorter (370 ± 29 msec) compared to baseline (388 ± 32 msec, $p < 0.001$) and standing (383 ± 34 msec, $p = 0.002$). QTc reached baseline values after 1 minute of recovery and remained stable thereafter (428 ± 19 msec at 1 minute, 434 ± 21 msec at 5 minutes, $p = 1.0$).
Conclusion: Significant QTc changes occur in healthy children in response to a standing test, yet with a greater magnitude than reported in adult series. This test needs to be validated in larger pediatric cohorts before its' clinical use.

P2037 - USEFULNESS OF ELECTROCARDIOGRAM IN THE DETECTION OF VENTRICULAR HYPERTROPHY IN ASYMPTOMATIC NEWBORNS

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Background: Historically, electrocardiogram (ECG) has been a key diagnostic tool in the work-up of congenital heart disease (CHD). Studies published in the past have helped establish the normal range for QRS amplitude and interval duration for newborns. At our hospital, we use ECG to screen infants of diabetic mother and newborns with a heart murmur. All newborns with ECG demonstrating ventricular hypertrophy receive an echocardiogram. Our objective is to determine the yield of echocardiogram performed in newborns with ECG demonstrating ventricular hypertrophy.
Methods: A retrospective chart review of infants born at our hospital, a tertiary care center, between 2013 to 2016 was performed. Of these, 777 full term, asymptomatic newborns had cardiac evaluation including echocardiogram. The ECG findings were reviewed and categorized into 2 groups: Group 1, with ventricular hypertrophy and Group 2, with no hypertrophy (Table 1). Echocardiogram findings were reviewed and divided into 4 categories (Table 2). Fisher's Exact test was used to compare the two groups.
Results: 676 infants had ECG performed as part of their work-up (Figure 1). Of these, 43 (6.4%) newborns had ventricular hypertrophy by echocardiogram. In Group 1, 25/234 (10.7%) newborns had hypertrophy by echocardiogram. Critical CHD was identified in 3 of these 25 newborns. Of note, 2 patients with Critical CHD only had an innocent murmur and echocardiogram was performed solely for ventricular hypertrophy detected on ECG. In Group 2, 18/442 (4.1%) newborns had ventricular hypertrophy by echocardiogram. There was a statistically significant difference in the incidence of ventricular hypertrophy identified between the 2 groups ($p < 0.02$).
Conclusion: ECG is an inexpensive and non-invasive tool to identify ventricular hypertrophy in newborns, although the yield is low. Further research is needed to redefine the voltage criteria and improve the specificity. ECG remains a critical tool in the identification of Critical CHD.

Table 1. Categorization of Bectrocardiogram Findings

Group 1: Ventricular Hypertrophy on ECG (n = 234)	Right ventricular hypertrophy
	Left ventricular hypertrophy
	Septal hypertrophy
	Biventricular hypertrophy
Group 2: No hypertrophy on ECG (n = 442)	Normal sinus rhythm
	Sinus bradycardia or tachycardia
	Non-specific ST-T wave changes, Prolonged QTc
	Axis deviation: right or left
	Premature atrial contraction.
	Premature ventricular contraction
	Atrial flutter. Supraventricular tachycardia

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Table 2. Categorization of Congenital Heart Disease (CHD)

Critical	any CHO requiring immedete htervention Example: critical aortic or pulmonary stenosis, hypoplastic left heart syndrome, transposition of the great arteries
Major	any CHD requiring interventbn within 6 months of life Example: large VSD, atrioventricular septal defect, tetralogy of faItot
Minor	any CHO expected to resolve spontaneously or unlikely to require intervention Example: small PDA, ASD or small muscular VSD.
Normal	No CHD Example: PFO, very small PDA and normal variants

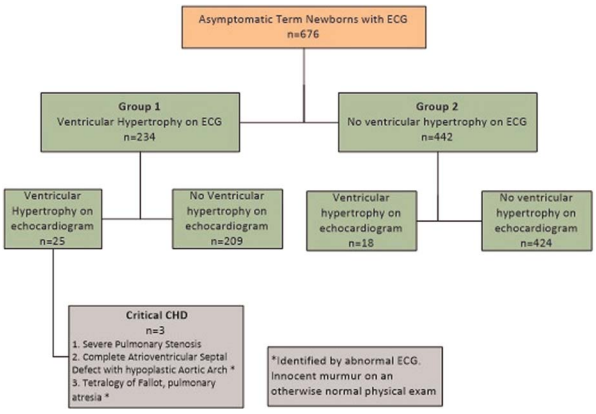

Figure 1.
Flow Chart Depicting Newborn Electrocardiogram Findings

P2052 - HEMODYNAMIC FACTORS INFLUENCING UPON EFFICIENCY OF ANTIARRHYTHMIC THERAPY IN INFANTS, TODDLERS AND PRESCHOOL CHILDREN

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Aim: To analyze the results of antiarrhythmic therapy (AAT) in preschool children with arrhythmias.
Methods and materials: 100 patients with arrhythmias got prolonged AAT. Arrhythmias were WPW syndrome, atrial tachycardias, ventricular tachycardias, extrasystole. Children's age was till one year old – 41 pts, from 1 till 3 - 23, from 3 till 7 - 36. Reasons for drug therapy were heart attacks prevention, sinus rhythm recovery or heart rate control at the level of normosystole with permanent and incessant tachycardias; depression and decrease of ectopic activity of arrhythmia focus. The therapy duration varied from 10 days till 12 months. The most children were prescribed monotherapy. When it was ineffective, in 19 patients, they got combined treatment. With the aim of assessment of Holter and EchoCG during prolonged AAT their comparison analysis was carried out initially, at 5-8 day after efficiency criteria appearance, and also in 6 months after therapy withdrawal.
Results: Criteria of effective therapy were reached in 38% of pts. Prolonged AAT was ineffective in 62% of children. The most effectiveness was marked with prolonged amiodarone intake in our patients. The drug was effective in 40,6% of patients, where the children with WPW syndrome at the age of under one year old predominated. The assessment of results of antiarrhythmic therapy using discriminative analysis allows defining factors influencing on its effectiveness. The factors were age and echocardiographic signs of tachyinduced cardiomyopathy (TCMP). Children under one year old with WPW syndrome, paroxysmal tachycardia and without TICM signs have greater chance of effective AAT.
Conclusion: Patients of early age with tachyarrhythmias which aren't accompanied by heart structural changes should expect effective result of drug therapy. Patients after one year old with echocardiographic signs of tachyinduced cardiomyopathy should expect ineffective AAT result and perform RFA.

P2054 - AMIODARONE-INDUCED THYREOPATHIES IN SMALL CHILDREN WITH ARRHYTHMIAS

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Aim: The aim of the study was to assess the thyroid hormone levels during prolonged amiodarone therapy in 0–7-year-old children with arrhythmias.
Materials and Methods: A total of 42 patients with WPW syndrome (n = 19), atrial tachycardias (n = 16), and ventricular tachycardias (n = 7) received amiodarone therapy. The duration of amiodarone administration varied from 0.7 to 24 months (Me 6.00; IQR 1.83-9.00). The thyroid hormone levels were estimated at the following time points: before treatment, during the amiodarone treatment at least 3 weeks after the beginning of therapy, and 6 months after amiodarone discontinuation.
Results: The levels of thyroid-stimulating hormone (TSH), total thyroxine, free thyroxine, thyroglobulin increased during amiodarone therapy.
The increases in the total and free thyroxine levels were statistically significant whereas TSH and thyroglobulin increased insignificantly during amiodarone therapy. All these indices significantly decreased 6 months after discontinuation of amiodarone therapy. Comparison of the initial values with the tests results 6 months after amiodarone discontinuation did not show any statistically significant differences. No clinical sings of hypertyrosinemia were observed in our patients during amiodarone treatment. The most significant changes in the levels of thyroid hormones were found in infants. During the entire course of amiodarone treatment, the median values of thyroid hormones did not exceed the upper reference ranges even when statistically significant changes in hormonal status occurred.
Conclusions: Amiodarone-induced changes in thyroid status were reversible and thyroid hormone values normalized 6 months after discontinuation of therapy.

P2139 - FASCICULAR VENTRICULAR TACHYCARDIA CASE REPORT

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Background: The complaint of palpitation in children is a symptom that should be carefully evaluated, especially if it is associated with symptoms of low cardiac output, since it allows adequate diagnosis and treatment, as well as orientation on the prognosis of the patient, depending on the arrhythmia identified.
Case Description: The purpose of this paper is to describe a case of a 11-year-old black male patient with no structural heart disease, with a history of palpitation associated with dizziness and pre-syncope since pre-school age. He was submitted to a Holter and was diagnosed with idiopathic fascicular ventricular tachycardia at the age of 5 years old. Since then, he has made regular use of verapamil. Due to partial symptom control, metoprolol was associated to the treatment. However, there was worsening of dizziness and fatigue, which improved with reduction of the beta-blocker dosage. Ergometric stress test showed no exercise tolerance. Transthoracic echocardiography revealed normal cavity dimensions, without global and segmental contractility alterations and ejection fraction of 54% by the Simpson method.
Discussion: Idiopathic fascicular ventricular tachycardia of the left ventricle is a rare manifestation of arrhythmia. Although it is a ventricular tachycardia, its origin usually occurs in the fascicles of the left branch, presenting a narrower QRS. It usually affects young patients without structural heart disease. It is characterized by its reversibility with the use of intravenous verapamil. Radiofrequency ablation is indicated for the treatment of symptomatic patients, since there is a high rate of success of the procedure, resulting in a life free of adverse events, with a prognosis equal to that of the general population.

P2154 - EVALUATION OF SUSTAINED TACHARRYHTMIAS IN PATIENTS YOUNGER THAN 1 YEAR OF AGE 6 YEARS OF EXPERIENCE IN SINGLE CENTER

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Background: Tachyarrhytmias are common cardiac arryhtmias in newborns and infants. In this study, we aim to evaluate the results of non-operational sustained tachyarrhytmias in patients younger than 1 year of age in our center.
Materials and Methods: The study was retrospectively performed in our center between November 2010 and November 2016 Post-operative tacharrhytmias were excluded. Demographical characteristics, type and localization of the tachyarryhtmia, echocardiographic findings and applied medical and/or ablation therapies were evaluated.
Results: In total we had 92 patients (60 male, 32 female). 86 had supraventricular tachcardia (SVT), 6 had Ventricular tachcardia (VT). Median age was 60 days (1-360 days), Median weight was 5 kg (2-10 kg). Common symptoms were palpitation and discomfort (n = 46, 50%) Others were detected during routine inspection (n = 29, 31%) and fetal echocardiography (n = 9,9.7%). 16 of them (18%) had left ventricular dysfunction in first echocardiographies, 9 had congenital heart diseases.The subtypes of SVT were determined in 55 (59%) of the SVTs by both invasive and non-invasive methods. 27 of them had WPW, 11 had FAT, 7 had concealed accessory pathway, 6 had PJRT, 4 had atrial flutter and the remaining 31 had SVT with narrow QRS. 83 of the SVT patients were treated using medical therapy. 9 cases were treated with ablation due to either resistance to medical therapy or left ventricular dysfunction. Commonly used medications were adenosine (all the patients), propranolol (n:70), amiodarone (n:31), flecainide (n:16), propafenone (n:15), esmolol (n:12), digoxin (n:14), sotalol (n:8). 11 cases had medical therapy combined with cardioversion.
Conclusion: Sustained tacharryhtmias in children under 1 year of age are mostly caused of accessory pathways and need multidrug therapy. It should be kept in mind that ablation is an effective alternative therapy in those cases with resistance to medical therapy and/or left ventricular dysfunction.

P2155 - SINGLE CENTER POPULATION BASED RESULTS FOR RADIOFREQUENCY CATHETER ABLATION OF TACHYARRHYTHMIAS IN CHILDREN

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Background: Tachyarrhythmia's are common in pediatric population and catheter ablation has become the definite treatment. The purpose of our study is to examine all catheter ablations performed in pediatric patients in our electrophysiology lab.
Methods: From January 2004 to November 2016, 274 children with tachyarrhythmia were submitted to electrophysiology study. Retrospective analysis of clinical features, electrophysiology study and follow up were performed. The median age (IQR) was 13 years (11-14), 49% female, median weight (IQR) 52 kg (43-64). 94% (257/274) of the patients received antiarrhythmic therapy, 96% (246/257) β-blockers, 18% (46/257) flecainide and 6% (15/257) others. Patients were followed for at least 1 year.
Results: 288 electrophysiology studies were performed in 274 patients, 28 (10%) were normal. Ablation was performed in 82% (215/260) of the studies. We found 176 accessory pathway in 163 patients, 56% (98/176) were left. Atrioventricular nodal reentrant tachycardia (AVRT) was present in 60 (20%) studies, ventricular tachycardia (VT) in 12 (4%) and atrial reentrant tachycardia (ART) in 7 (2%). Global RF ablation success was 94% (203/215). Accessory pathway were successful overall in 94% (142/151) of ablations, 99% (94/95) in left and 86% (48/56) in right ($p = 0.003$ Mantel-Haenszel with Yates correction). AVRT ablation was successful in 96% (46/48), VT in 55% (6/11) and ART in 80% (4/5). We had 8 (3%) minor complications: transitory atrioventricular block, arteriovenous fistula and femoral hematoma. No major complication was reported. The overall recurrence rate was 21/203 (10%), 11% in accessory pathway and 4% in AVRT.
Conclusions: The RF ablation is a safe and effective treatment for pediatric patients with tachyarrhythmia. In our cohort we had similar overall ablation success and recurrence rate as reported in literature.

P2157 - CATHETER ABLATION OF MANIFEST POSTEROSEPTAL ACCESSORY PATHWAY ASSOCIATED WITH CORONARY SINUS DIVERTICULA IN A CHILD WITH CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

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Background: To date, a pediatric patient with congenitally corrected transposition of the great arteries (CCTGA) accompanied by WPW syndrome treated by an ablation inside the coronary sinus (CS) diverticula has not been reported. This study presents a successful radiofrequency (RF) catheter ablation of the CS diverticula–sourced manifest accessory pathway in patient who was diagnosed with CCTGA.
Case Report: A 4.5year-old male patient, weighing 18 kg, visited our clinic with a repetitive multiple antiarrhythmic-resistant (including beta-blocker + propafenone + amiodarone) supraventricular tachycardia (SVT). The patient had a pulmonary banding operation when he was 2 months old because of CCTGA, broad perimembranous VSD, right ventricle hypoplasia, and pulmonary hypertension. Also he had a two successful RF catheter ablation operation because of the left posteroseptal region–related AP (WPW), but SVT was recurred. After discussing possible risks with his family, he underwent catheter ablation again. An electrophysiological study was performed after an informed consent was obtained. Considering the patient's age and complex cardiac pathology, EnSite Velocity System (St. Jude Medical Inc.,MN, USA) and fluoroscopy were used during ablation operations. During the electrophysiological study as the earliest area was found as CS ostium, a mapping was done into the CS, and CS diverticula were detected with the help of CS angiogram. Here, the place that was 40 ms ahead of the surface delta was labeled with an RF catheter mapping. To be able to detect the proximity between coronary artery and ablation area, a coronary angiography was performed. Since the ablation area was far from the coronary arteries, RF ablation was performed successfully. The patient was asymptomatic without any drug during the 9-month follow-up, and no preexisting transition was observed on ECG.
Conclusion: Manifest AP ablation can be difficult in congenital heart diseases such as CCTGA. In addition, CS diverticula should be taken into account in recurrent posteroseptal APs.

P2158 - ATRIAL FLUTTER AND NONCONDUCTED BIGEMINY PREMATURE ATRIAL CONTRACTION IN A NEONATE WITH CARDIAC RHABDOMYOMA

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Background: In the present case study, an 8-day-old neonate whose bigeminy nonconducted premature atrial early pulse–related bradycardia and cardiac rhabdomyoma–related resistant atrial flutter were detected and successfully treated was presented.
Case Report: This study presents the case of a baby with birth weight 3100 g born after an unfollowed antenatal pregnancy and visited another medical center with a decrease in the suckling reflex and cyanosis complaints. In the physical examination, tachycardia with an irregular (with a narrow QRS phase) RR interval (up to 200/min) was detected in the baby. In the 2D and color Doppler echocardiographic evaluation, a slight hyperechogenic rhabdomyoma (13 × 15 mm2) near the superior vena cava at atrium fornix was observed in the subcostal apical five-chamber section (left side). It was observed that the tumors did not cause any obstruction in the flow, and the functions of the left ventricle was conserved. Despite these medical treatments, the patient's atrial flutter continued; therefore, the atrial flutter was stopped with cardioversion, which was synchronized with 2 J/kg energy. A frequent-aberrant transitive and bigeminy nonconducted premature atrial contraction (PAC) related bradycardia was detected in the after-cardioversion follow-ups of the patient, and the treatment was carried out after giving oral 3 mg/(kg · day) amiodarone. The atrial flutter and bradycardia attacks of the patient did not repeat during the 3-month follow-up. This patient was still followed up for tuberous sclerosis and was under low-dose amiodarone + propafenone treatment.
Conclusion: In neonates, rhabdomyomas with atrial location can cause problems both by making atrial flutter with fast ventricle transportation and by leading to bigeminy nonconducted PAC-related functional bradycardia. Medical treatment and synchronized cardioversion are usually seen enough to treat that kind of arrhythmia.

P2169 - RADIOFREQUENCY ABLATION OF FAST VENTRICULAR TACHYCARDIA THAT CAUSED AN ICD STORM IN AN INFANT WITH HYPERTROPHIC CARDIOMYOPATHY

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Background: An implantable cardioverter defibrillator (ICD) storm involves very frequent arrhythmia episodes and ICD shocks and is associated with poor prognosis in both the short and long term. The radiofrequency catheter ablation (RFCA) process can be used as a valuable salvage treatment for patients with an ICD storm.
Case Report: A two-year-old male patient weighing 12 kg was transferred to our hospital after cardiac arrest (CA) caused by fast VT. Defibrillation was repeatedly applied. The patient's echocardiography revealed non-obstructive hypertrophic cardiomyopathy (HCM). We carried out a propranolol, amiodarone, and flecainide treatment, and for secondary prevention the patient received an epicardial ICD. During the followup, "incessant" fast VT episodes of distorting hemodynamics, ICD shocks, impairment of systolic functions, and development of hypotension occurred; the patient was intubated and moved to the intensive care unit. Despite the intensive medical therapy and antitachycardia pacing, the treatment was ineffective, the VT attacks and ICDshs could not be stopped—more than 50 occurred in one day. The patient was then taken to the catheterization room for electrophysiology study (EPS) and ablation, and then in intubated condition from the ICU with intravenous support treatments and extracorporeal life support (ECMO) to the EPS laboratory. A successfully RFCA was performed salvage treatment for the fast VT that left posterior fascicular's Purkinje fibers and RF "line" lesions were created in the Purkinje area up to the left posterior fascicle(LPF)-HIS connection. After the ablation, the patient was transferred in a stable condition to the ICU with no need for an ECMO. During the six months follow-up after the antiarrhythmic treatment, the patient experienced no more ICDshs.
Conclusion: To our knowledge, this is the first infant case among HCM-diagnosed children with an ICD storm on whom an radiofrequency catheter ablation salvage treatment for the fast VT.

P2183 - IS THERE A CORRELATION BETWEEN ELECTROCARDIOGRAPHIC P WAVE FEATURES AND GOSE INDEX IN EBSTEIN'S ANOMALY

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Background: The aim of the study is to investigate the correlation between GOSE index of the patients with Ebstein's Anomaly and their electrocardiographic p wave amplitude and dispersion.
Patients and Methods: Unoperated patients with Ebstein's Anomaly were included prospectively in the study. The echocardiographic GOSE indexes and the simultaneous ECGs of the patients were recorded. The echocardiographic calculations and ECG evaluations were performed by the same pediatric cardiologist.
Results: 24 patients were evaluated in this study. Twelve of the patients were female (50%). The median body weight was 15 kg (2,5-60 kg), median O2 saturation was 95% (75-100%), median GOSE index was 0,62 (0,2-1,3), median p wave dispersion was 18msec (8-38msec), P max was 86 msec (58-104), P min was 62 msec (18-88) ve P amplitude was 2.7 mm (1.2-5). GOSE index was $<0,5$ in 8 cases, between 0,5-1 in 12 cases and >1 in 4 of the cases. A positive relation was observed between the GOSE index, P max ($r=0.5$, $p=0.02$) ve P amplitude ($r=0.780$, $p=0.001$).
Conclusion: There appears to be a significant correlation between GOSE index which is important in determining the severity and prognosis of the Ebstein's patients and p wave amplitude. Gose index might be predicted by the amplitude of p wave. For the confirmation of the importance of ECG data in prognosis, larger series of patients are needed to be evaluated.

P2191 - A CASE REPORT IS MEXILETINE USAGE EFFECTIVE IN SHORTENING THE QTC DURATION AND IMPROVING T WAVE ALTERNANS IN TIMOTHY SYNDROME

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Background: This article aimed to present a case with Timothy syndrome (TS) with cardiac arrest history, prolonged QT, and T wave alternans. TS is a rare multisystemic disease characterized by facial deformation, malign ventricular arrhythmia, syndactyly, immune deficiency, intermittent hypoglycemia, autism, neurocognitive development retardation, and prolonged QT duration (1). After mexiletine administration, the QTc duration of the patient got shortened and her T wave alternans disappeared.
Case Report: A 4.5-year-old female patient, who had a cardiac arrest history, presented with several Torsades de Pointes and ventricular fibrillation attacks during her intensive care follow-ups in another hospital. This patient was directed to our center after seeing a prolonged QTc in her electrocardiograms (ECGs). She had syndactyly on each foot. All other systemic examinations were normal except hypertonicity and contracture that occurred secondary to previous hypoxia. The ECG at arrival was at a normal sinus rhythm, with a heart rate of 90/min and QTc value of 606 ms. In her Holter ECG records, her QTc interval was extremely long (>600 ms), and she had a T-wave alternans. A positive Pg406R mutation in CACNA1C gene was found which is specific in TS. An epicardial implantable cardioverter defibrillator was implanted to the patient, and propranolol treatment was started. However, propranolol alone was not adequate to reduce the heart rate or to improve the T wave alternans. An additional mexiletine treatment was started. In her consecutive electrocardiograms, her heart rate decreased to 78/min, QTc value decreased to 501 ms, and T-wave alternans disappeared.
Conclusion: This case presents particular importance to the authors as it bandies about the idea of mexiletine usage in addition to an initial beta-blocker treatment for preventing T-wave alternans and decreasing the QTc interval in the cases with TS.

P2260 - VENTRICULAR TACHYCARDIA DUE TO NKX2.5 MUTATION A FIRST CASE REPORT

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Background: The phenotype of familial arrhythmia syndromes is expanding with the increased use of genetic testing, robust cascade testing as well as the improved ability of implanted devices to recognize different arrhythmias. NKX 2.5 mutations have traditionally been associated with septal defects and cardiac conduction disorders. Recently, the possibility of association of NKX2.5 mutation with ventricular arrhythmias was raised based on animal studies and clinical hypothesis based on one report. We report a child with history of sudden death and AV conduction disorder with a mutation in NKX2.5 who was detected to have a ventricular arrhythmia on pacemaker and was uneventfully upgraded to an implantable cardioverter defibrillator.
Methods: This was a retrospective case record review of a family with history of septal defects, cardiac conduction disorders and sudden cardiac death.
Results: A 15-year-old boy was evaluated because of family history of sudden cardiac death. His father had underwent surgical closure of atrial septal defect in childhood and had been implanted with a permanent pacemaker after atrial fibrillation as diagnosed at 20 years of age. He died suddenly at 35 years of age and rapid ventricular beats prior to death was detected on the pacemaker. His brother had also undergone surgical closure of ASD at 3 years and had died suddenly at 18 years of age. The boy had also undergone ASD closure in early childhood. High degree AV block was detected on Holter and he underwent a pacemaker implantation. Routine pacemaker interrogation revealed 2 episodes of non-sustained ventricular tachycardia. He was upgraded to an implantable cardioverter defibrillator. Genetic testing revealed a causative mutation in NKX 2.5 gene. Family screening revealed evidence of cardiac conduction disorder in his half-sister.
Conclusion: NKX 2.5 mutations can cause ventricular arrhythmias in addition to A-V conduction disorders and a pacemaker may not be life saving in all.

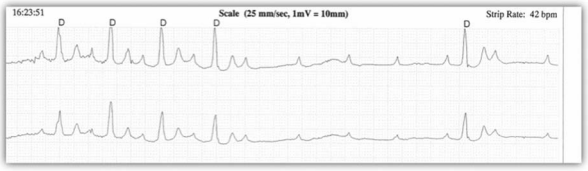

Figure 1.

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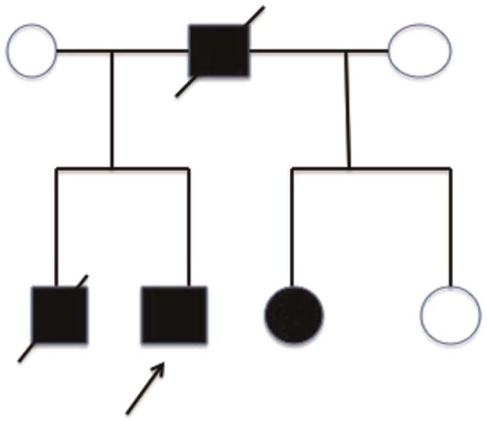

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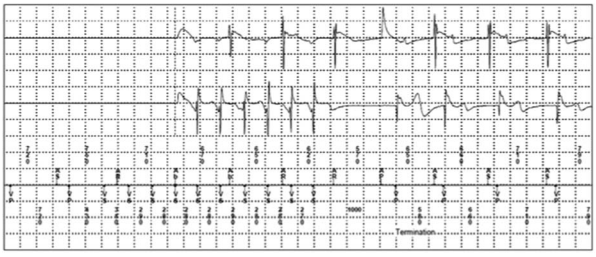

Figure 3.

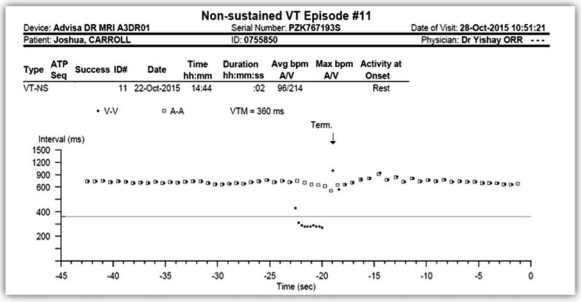

Figure 4.

P2302 - EFFECT OF HAART (HIGHLY ACTIVE ANTIRETROVIRAL TREATMENT) ON QTC INTERVAL IN ANTIRETROVIRAL NAÏVE HIV INFECTED CHILDREN A PROSPECTIVE LONGITUDINAL STUDY

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Background: Prolongation of QTc interval, a complication which can lead to serious arrhythmia or sudden death, has been well documented in HIV-infected adult. However, there were limited studies in children with conflicting results regarding cause and associated factors.
Objective: To determine the prevalence of prolonged QTc interval and factors associated with longitudinal change after HAART regimen in antiretroviral naïve HIV-infected children.
Method: Prospective longitudinal study of 12-lead EKG in 40 antiretroviral naïve HIV infected children who were enrolled to received HAART at pediatric infectious clinic, Chiang Mai University Hospital, Thailand. The QTc interval (Bazett's formula) were measured at baseline, 1 year and 2 years after HAART regimen. Linear mixed model analysis was used to analysis factors associated with QTc changes over 2 years of treatment.
Result: Of 40 asymptomatic HIV-infected children enrolled, 32 patients (80%) were assigned to efavirenz based regimen and 8 patients (20%) were assigned to navirapine based regimen. There were 35% male and the mean age was 7.8 ± 2.6 years. Median CD4 percentage at baseline was 3% (interquartile range 1-8%) and mean viral load was 5.37 ± 0.38 log10copies/ml. Before initiation of HAART, the mean QTc interval was 409 ± 22 msec and no patient had QTc prolongation. After HAART, mean QTc interval was significant longer to 420 ± 24 msec at 1 year ($p = 0.006$) and 421 ± 27 msec at 2 year ($p = 0.006$). Factors associated with longitudinal increased in QTc interval were increasing age (estimate 1.86, 95% CI 0.52-3.18, $p = 007$) and increasing LDL-cholesterol (estimate 0.16, 95% CI 0.03-0.28, $p = 0.014$).
Conclusion: Asymptomatic antiretroviral naïve HIV-infected children had no QTc prolongation. After HAART regimen, increasing age and LDL-cholesterol level were associated with QTc prolongation.

P2322 - SUCCESFULL RADIOFREQUENCE ABLATION OF ECTOPIC FOCI IN A TODDLER WITH ECTOPIC ATRIAL TACHYCARDIA AND AMIODARONE ANAPHYLAXIS

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Background: Ectopic atrial tachycardia (EAT) is a common chronic supraventricular tachycardia (SVT) in children that often resists medical therapy. Untreated, it may progress to tachycardia-induced cardiomyopathy. It is often resistant to medical therapy and radiofrequency ablation is a succesfull and safe therapy option. Amiodarone is a class III antiarrhythmic drug, frequently used and very effective in ventricular and supraventricular arrhythmias; however, it has numerous side-effects, involving many organ systems. However anaphylactic reaction to amiodarone is a very rare caomplication.
Case Report: A two years old girl was referred to our center due to SVT resistant to adenosine. The electrocardiogram revealed a long R-P narrow QRS tachycardia, with blocked p waves under adenosine effect consistent with EAT. Echocardiography revealed only patent formanen ovale with normal left ventricular systolic function. During amiodarone infusion in PICU diffuse maculopapular rash and respiratory distress evolved, so amiodaron infusion was stopped and steroid and antihistaminics were given. The signs dissolved rapidly and class II (propranolo) and classI (flecainid) agents were started. On ambulatory follow-ups recurrent and incessent tachcardia attacks repeated and a class III agent (sotalol) was added to the therapy. Due to incessent tachcardias after multidrug therapy with 3 agents, an electrophysiologic study with successfull radiofrequency (RF) ablation of the ectopic foci was conducted. Conventional and ESV system mapping during tachycardia revealed two different foci in the right atrial appendage. The foci were marked with ESV system and ablated with a 5 F RF catheter successfully. The procedure was accomplished without any complication. At the follow-ups, the patient reports no further episodes of tachycardia.

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Conclusion: In conclusion we want to evoke that antiarrhytmic drug therapy is often ineffective and complicated in EAT in children and radiofrequency catheter ablation of such cases is a safe and succesfull therapy modality, even in younger children.

P2368 - THE PARADOX OF BRADYCARDIA AND WIDE QRS

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Background: We report the clinical presentation and management of two infants presenting with acute Flecainide toxicity without history forthcoming of medication administration error.
Methods: N/A
Results: Patient (1): 4-month-old male with d-transposition of the great arteries post arterial switch operation and history of atrial tachycardia managed with Flecainide 6 mg/kg/day and Propranolol 3 mg/kg/day. Patient (Pt) presented with 2-day history of decreased activity, lethargy, and electrocardiogram (ECG) showing sinus bradycardia (rate 66 bpm), first degree atrioventricular (AV) delay (PR 92 ms), severe intra-ventricular conduction delay (IVCD, QRS 160 ms – 242 ms), and premature atrial contractions conducted with variable rate-related bundle branch aberrancy (RRBBA). Figure shows representative ECG. Patient (2): 4-week-old female infant with reentrant supraventricular tachycardia managed on Propranolol 5 mg/kg/day and Flecainide 7 mg/kg/day. Pt presented with 2-day history of lethargy, poor oral intake, and ECG showing sinus rhythm with first degree AV delay (PR 206 ms), severe IVCD (QRS 180 ms), and premature atrial complexes conducted with variable RRBBA. In each case, diagnosis of Flecainide toxicity was suspected based on the ECG findings involving first degree AV delay and severe IVCD. High dose intravenous sodium bicarbonate was initiated along with supportive management with resolution of electrocardiographic findings in approximately two days in each case. Admission serum Flecainide confirmed toxic levels (Pt 1: 1.73 mcg/mL; Pt 2: 2.6 mcg/mL; therapeutic range 0.2-1.2). Pt 2 was discharged on optimized dose of Propranolol and Pt 1 was restarted on an appropriate dose 3 weeks post-discharge due to recurrence of SVT, with no further recurrences documented during their follow-up.
Conclusion: Flecainide toxicity remains an important complication seen during management of tachyarrhythmias. Prompt recognition of ECG manifestations and discontinuation of the drug along with institution of high dose intravenous sodium bicarbonate aids in the reversal of ECG manifestations and recovery.

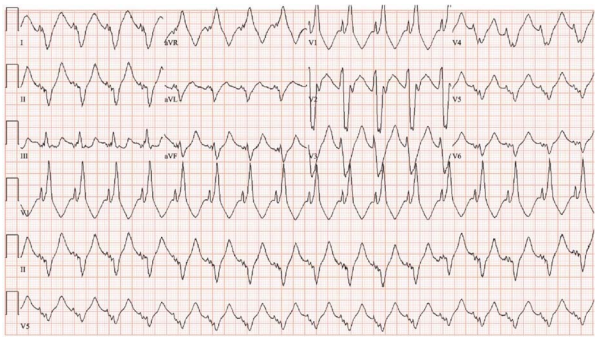

Figure 1.

P2410 - THE MEASUREMENT OF QTC IN CHILDREN A SURVEY OF CLINICAL PRACTICE

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Background: The QT interval is key to the diagnosis of Long QT Syndrome. Accurate and reproducible measurement of the QT interval can be challenging in clinical practice.
Methods: We conducted a questionnaire based survey to evaluate the methodology used in practice in the UK.
Results: The survey of 109 clinicians consisted of Paediatric electrophysiologists (7), Paediatric cardiologists (32), and trainees (27) as well as pediatricians with expertise in cardiology (24) and trainees (19). All participants used Bazett's formula for rate correction. Only 10.1% used additional formulae (7 Fredericia's, 2 Framingham and 2 QT nomogram). 83.3% measured the RR interval preceding the QT interval, 13.9% the concurrent RR interval and 0.9% RR unrelated intervals. 15.7% used manual calipers and 7.4% used electronic calipers for measurement. When intervals were estimated visually (without electronic calipers), the nearest approximation aimed for during measurement varied- 40msec 19.8%, 20msec 35.4%, 10msec 42.7% and 4msec 2.1%. 82.6% preferred lead II or V5 for measurement while the rest used a range of other chest and limb leads as their preferred choice. Of those using multiple cardiac cycles for measurement 50.6% quoted the average, 48.1% quoted the longest QTc and one quoted the range. 50.5% used the 'tangent method' to determine the end of T wave whilst 49.5% used visual assessment. If U-waves were present, 46.8% would always ignore it. 27.5% included U waves that merged with T-waves and 13.8% included U-waves that were at least half the T-waves height. The findings were broadly similar between the clinician categories.
Conclusion: There is no uniformity in the methodology of QTc measurement among clinicians in the UK. This study highlights the need for a nationwide consensus on methodology to ensure accurate and reproducible measurements.

P2412 - IMPROVED SIGNAL RESOLUTION AND FEWER LESION ATTEMPTS USING A MICROFIDELITY ABLATION CATHETER IN PEDIATRIC WPW

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Introduction: The Intellatip MiFi ablation catheter is an 8mm RF ablation catheter (Boston Scientific) with three circumferentially located microelectrodes at the catheter tip used to provide high fidelity local electrograms (EGMs) for mapping and identification of optimal ablation sites. We hypothesized that the MiFi catheter has utility in pediatric WPW ablation with improved signal resolution to identify accessory pathway (AP) potentials and lead to shorter procedures, reduced fluoroscopy, and fewer lesions.
Methods: Retrospective review of pediatric manifest WPW ablations at our center from Dec, 2012 to Jan, 2017. For safety with 8mm RF ablation, the MiFi catheter was only used in patients >35 Kg and with shorter RF application times. There were 38 cases with standard ablation catheters (Blazer, Boston Scientific, 4/8 mm) and 25 cases with MiFi. In 4 excluded cases, the MiFi catheter was successfully used for mapping only to identify ideal cryoablation site for parahisian AP.
Results: Long term success with at least 1 month follow up was achieved in 36/38 standard (std) cases and 25/25 MiFi cases. First lesion success was achieved in 8/38 (21%) std cases and 14/25 (56%) MiFi ($p < 0.05$). MiFi cases had fewer total lesions (avg 4.3 vs. 11.4;$p < 0.05$). In most MiFi cases (16/25), the microfidelity

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EGMs demonstrated a clear, sharp, and higher amplitude AP potential (Kent potential) compared to standard ablation distal bipole. Avg total procedure time was not significant, while avg fluoro time (0.04 min MiFi vs. 1.1 min; p = 0.06), and radiation dose (0.45 mGy MiFi vs. 8.12 mGy;p = 0.07) trended toward significance. The intent was to remain fluoroless in all cases. No complications in either group.
Conclusion: These data suggest that the MiFi catheter has benefits over standard ablation mapping for identification of AP potentials in pediatric patients with WPW and allows for fewer total RF lesions with reduced fluoroscopy time.

P2606 - TRANSSEPTAL CATHETER APPROACH FOR LEFT SIDED ACCESSORY PATHWAYS – THE "GENTLE WAY" OF ABLATION FOR CHILDREN AND ADOLESCENTS

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Background: Radiofrequency catheter ablation (RFCA) of accessory pathways (APs) is the treatment of choice for AP-mediated supraventricular tachycardias. Since its introduction in children using the transaortic approach via retrograde femoral arterial cannulation for left-sided APs there were concerns regarding mechanical vascular and valvular damage. Thanks to extensive experience with transseptal (TS) punctures in adults this approach is now frequently used. However, the "conventional" retrograde transaortic approach continues to be the preferred choice in children.
Aim: This retrospective single-center analysis is focused on assessment of safety and efficacy of transseptal approach for RFCA of left-sided APs in children.
Material and Methods: In the period of 9 years (1/1/2008–31/12/2016) 67 ablations of left-sided APs via TS approach were performed in 65 children and TS approach was used in additional 11 pts for mapping (AP was ablated at the right side eventually). TS puncture was necessary in 59 pts (75,6%), the remaining 19 had a patent foramen ovale. For TS puncture long fixed-curve or variable curve sheaths were used (SL1 or Agilis, St. Jude Medical, USA) together with the corresponding TS Brockenbrough needles (BRK, SJM). I.V. heparin was administered after the puncture and repeated after 45 minutes.
Results: 59 TS punctures were performed in 57 children (6–18 years old, median 15). All procedures were successful without complications. RFCA was acutely successful in 55/57 pts with successful reablation in 2 patients and clinical long-term elimination of the arrhythmia in all but 1 pt (98%).
Conclusion: RFCA of left-sided APs in children via TS approach represents in our experience a very safe and effective ablation technique. We believe that TS approach for RFCA of left-sided APs eliminates any concerns of damage to subtle vascular and valvular structures in children and should be the approach of choice for experienced operators.

P2617 - UNREPAIRED TETRALOGY OF FALLOT WITH MYOCARDIAL INFARCTION IN AN ADULT PATIENT

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A 66-year-old man with a history of unrepaired tetralogy of Fallot and diabetes mellitus presented to a community hospital for worsening dyspnea and orthopnea. On admission, the patient was in New York Heart Association functional capacity III and continuous infusion of diuretics was administrated. After the admission, his heart rhythm changed from sinus rhythm to junctional rhythm and immediately after the change asystole and subsequent ventricular fibrillation were documented. Ventricular fibrillation terminated spontaneously and then the patient was transferred to our institution for further management. Physical examination was notable for a III/VI systolic ejection murmur and clubbing of the fingers was noted. Blood pressure was 148/84 mmHg and peripheral oxygen saturation 80% in air. Electrocardiography showed first-degree atrioventricular block with a cardiac rate of 70 bpm, rightward axis with right bundle branch block, and prolonged QRS duration (200 msec). Echocardiography revealed wide ventricular septal defect, with over-riding of the aorta, pulmonary valve stenosis with a peak gradient across the valve of 91 mmHg and right ventricular (RV) hypertrophy. The RV contractile function was severely impaired and left ventricular (LV) ejection fraction was decreased as well (30%). Moreover, left ventricular anterior and anteroseptum segments were hypokinesis. Coronary angiography showed 99% stenosis in the proximal left anterior descending artery and coronary stent implantation was underwent subsequently. After further management of heart failure, a cardiac magnetic resonance imaging demonstrated reduced RV and LV ejection fraction (24% and 21%, respectively). Due to the severe biventricular dysfunction, the option of total surgical repair was declined and cardiac resynchronization therapy with defibrillator (CRT-D) was considered. To predict the response of CRT, acute hemodynamic response was evaluated by invasive measurement of LV-dp/dt at baseline and biventricular pacing. LV-dp/dt increased from 720 mmHg/s to 940 mmHg/s after biventricular pacing and thus the patient underwent CRT-D implantation.

P2886 - CARDIAC TUMOUR MASQUERADING AS VENTRICULAR TACHYCARDIA

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Background: Cardiac tumours have been identified in 0.02% to 0.04% of the pediatric population. Most common of these tumours are benign. Large tumors may result in compression of cardiac chambers,conduction tissue or coronary vasculature, as well as obstruction of cardiac valves and outflow tracts. Some tumours display the potential for malignant arrhythmias, with an increased risk of sudden cardiac death. In the absence of hemodynamic impairment, observation is the standard care in most cases because many tumours spontaneously regress with time. Case reports have described the use of ICDs for persistent arrhythmias due to nonresectable tumours. We describe the case of a girl with a solitary large cardiac tumor, in the LV free wall, presenting with arrhythmias.
Materials and Methods: 12 year old Ms MD, presented with shortness of breath, NYHA II since early childhood. She had documented ventricular arrhythmia since the last two years, including prolonged episodes of monomorphic Ventricular Tachycardia

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(VT) at a ventricular rate of 150 beats/min to 175 beats/min as well as a slower wide complex rhythm, all of which were asymptomatic.Beta blockade with metoprolol was initiated with some improvement in the frequency. However, persistent arrhythmias prompted a conversion to sotalol. She continued to have asymptomatic episodes of VT, albeit less frequently. Echocardiogram revealed a 16 cm × 7 cm intramyocardial mass on the LV free wall.The mass seemed to be invading the LV cavity ,and was inhomogenous. Serial echocardiograms demonstrated rate related cardiac dysfunction. Cardiac CT scan demonstrated the nature of the tumor and its extent. Excision biopsy was contemplated,but only surgical biopsy was done due to the size and localisation of the tumor,with intraluminal extension, Histopathological specimen confirmed the tumor as a fibroma.
Results: The patient was followed with antiarrhythmic treatment. She has had a mild symptomatic improvement. *Conclusions:* Cardiac tumors, though rare in the general population,can present in a myriad masquerading ways. A good clinical judgement, along with proper imaging can be a saviour.

P2887 - CONCURRENT ANK2 AND RYR2 MUTATION IN A PATIENT WITH REFRACTORY CATECHOLAMINERGIC POLYMORPHIC VENTRICULAR TACHYCARDIA

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Backgourd: Inherited cardiac arrhythmia including long QT syndrome (LQTS)and catecholaminergic polymorphic ventricular tachycardia(CPVT)is an important cause of sudden cardiac death in young people.
Case Report: We report a rare case of young patient who developed refractory catecholaminergic polymorphic ventricular tachycardia (CPVT) with borderline QT interval and sinus bradycardia on resting state. He had seizure-like episodes from 15 months old, and put on beta blocker from 7 years old with diagnosis of CPVT. Because of frequent syncope, he received an implantable cardioverter-defibrillator. Nevertheless he experienced ICD storm and frequent ICD shocks , which was partly relieved by thoracoscopic left sympathectomy and flecainide. In molecular genetic testing, concurrent mutations of ANK2 for Long QT syndrome type 4 and RYR2 for CPVT were confirmed.
Conclusion: we demonstrate the simultaneous occurrence of both a putative pathogenic RyR2(Ryanodine receptor 2)mutation and ANK2(Ankyrin 2)mutation in patient with clinically overlapping refractory CPVT and LQTS4 phenotype for the first time in Korea. Further comprehensive functional analysis in vivo and vitro is positively necessary.

P3038 - IMPLANTABLE CARDIOVERTER DEFIBRILATORS IN CHILDREN AND PATIENTS WITH CONGENITAL HEART DISEASES WHAT HAVE WE LEARNT IN TWO DECADES OF FOLLOW UP

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Backgound: Implantable cardioverter defibrillators (ICDs) are a recognized therapy for the prevention of sudden death in adults. However, its use in children or patients with congenital heart disease (CHD) involves multiple drawbacks, and there is little information on the use and implant methodology. The aim of our study was to communicate the characteristics of the patients with ICDs from our center and to study the association between the presence of appropriate therapies and unfavorable events (electrode problems or inappropriate therapies) in both groups.
Materials and Methods: A retrospective, descriptive study.
Results: 46 patients (23% female). Average age of ICDs implant: 19 years (median 16 years). 20 patients with CHD and 26 with hereditary diseases (HD) (canalopathies or cardiomyopathies). 76% ICDs were implanted in secondary prevention. All but 5 were endocavitary systems; 1 epicardial, 2 mixed resynchronizing defibrillators and 2 with accessory subcutaneous coils. About chamber location: 10 single-chambered, 30 bicameral and 6 tricameral. Outcomes with mean follow up of 20 years were: 52% of patients had appropriate therapies, 28% inappropriate theraphies (23% due to supraventricular tachycardia and 5% due to electrode dysfunction). 10 patients presented electrode problems: 4 dislocations and 6 dysfunctions, more frecuent in younger patients. Five patients died, only one secundary to an arrhythmic storm. There was more incidence of appropriate therapies in patients with HD and, curiously, in those with ICDs as primary prevention. There was no difference by age. Inappropriate therapies were more frequent at lower ages, single-chamber devices, HD and ICDs implanted as secondary prevention.
Conclusions: The use of ICDs in children and CHD patients is of unquestionable utility. In our 20-year serie, ICDs have acted adequately in more than half of patients, but there are problems such as electrode dysfunction or inappropriate therapies, more frequent in lower ages.

P3046 - JUNCTIONAL ECTOPIC TACHYCARDIA IN CHILDREN EVALUATING TIME TO RATE CONTROL AND IMPACT ON OUTCOMES

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Background: Junctional ectopic tachycardia (JET) is the most common tachyarrhythmia in children after surgery for congenital cardiac disease (CHD). It is often self-limiting, but is associated with increased risk of morbidity and mortality. The contributing factors and impact of time taken to achieve rate control of JET is poorly described.
Methods: A retrospective, single-center cohort study of children who developed JET after CHD surgery from January 2010 to June 2015 was performed. We classified the cohort into 2 groups: patients who achieved rate control of JET in $\leq$24 hours and in >24 hours. We examined factors associated with time to rate control and compared clinical outcomes [mortality, duration of mechanical ventilation (MV), length of intensive care unit (ICU) and hospital stay] between the two groups.
Results: Our cohort included 27 children, with a median age of 3 months [interquartile range (IQR): 0.7-38]. The most common CHD lesions were ventricular septal defect (n = 10, 37%), tetralogy of Fallot (n = 7, 25.9%) and transposition of the great arteries (n = 4, 14.8%). 15 (56%) and 12 (44%) patients achieved rate control of JET in $\leq$24 hours and >24 hours respectively. There

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was a difference in median MV time [97 (21-145) vs 311 (100-676) hours, p = 0.013] and ICU stay [5.0 (2-8) vs 15.5 (5.5-32.8) days, p = 0.023] between the patients who achieved faster rate control than those who didn't. Differences in length of hospital stay and mortality between the 2 groups were not significant.
Conclusion: Our small cohort study demonstrated that time taken to achieve rate control of JET was associated with increased duration of MV and ICU stay, but did not impact mortality or length of hospital stay. Further larger studies are required to validate our preliminary findings.

HUMANITARIAN

P1014 - RESULTS OF INTERNATIONAL ASSISSTANCE FOR PEDIATRIC HEART SURGERY PROGRAM IN A SINGLE UKRAINIAN CENTER

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Background: Surgery for congenital heart disease has been slow to develop in parts of the former Soviet Union. We describe the impact of an 8-year surgical collaborative assistance program between our center and invited teams from countries with developed cardiac programs.
Material and Methods: The refered international pediatric assistance program included four main components: (1) on-site surgical and interventional activity with hands-on training of the local team, (2) medical education, (3) biomedical engineering support, 4) a team-based practice development. Data was analyzed from visiting teams and Kharkiv databases prior to and since commencement of assistance (Period A: 2000–2007 (8 y); Period B: 2008-2015 (8 y)). Out of a total of 1347 surgical and interventional pediatric cases performed during both periods, 979 surgical cases with RACHS-classifiable lesions were analyzed. We compared the following variables between periods: annual case volume, 30 day-hospital mortality, case complexity (RACHS-1), and RACHS-adjusted standardized mortality ratio (SMR: observed/expected mortality). We evaluated the improvement in local surgical skills during Period B.
Results: 154 RACHS-classifiable surgeries were performed during Period A, with a mean annual case volume of 19.3 surgeries (95% CI 14.3–24.2) an overall mortality of 4,6% and a SMR of 3.4. During Period B volume increased to 825 mean annual surgeries ($p < 0.0001$), with an increment to 103.1 mean annual cases (95% CI 69.1–137.2). The increase in case complexity led to a small raise of the overall mortality (5.7%) with a lower SMR (1.4). The proportion of local surgeon led surgeries during trips increased from 0% in 2008 to 98% in the final year of analysis.
Conclusions: This model of assistance to developing programs has improved adjusted mortality, increased case volume, increased complexity and developed independent operating skills in an economically disadvantaged center.

P1028 - PEDIATRIC CARDIAC SURGERY FOOTPRINT ITS GLOBAL IMPACT ON HUMAN DEVELOPMENT

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Objective: By using the concept of "Footprint of Global Surgery" we are trying to unveil the positive consequences of changing the natural history of congenital heart disease in low and middle income countries (LMIC). We attempt to measure the footprint of pediatric cardiac surgery in relationship to gains in a number of indicators (Life Expectancy, Education and Income per Capita) conforming the Human Development Index (HDI).
Material & Methods: We collected all clinical data available on patients operated by our organization in 10 LMIC during 2015. We also estimated from our financial data the operative costs incurred in the accomplishment of our missions during that year. Data on HDI indicators were obtained for each country while independent reviewers calculated the incremental difference in life expectancy, education and years of income per capita added by modifying the natural history of the disease by our intervention (surgical repair). The total financial cost of operations was divided by the number of patient reached with the global effort. A cost per surgery was achieved and ascribed to the incremental difference in those indicators.
Results: During 2015 a total of 470 operations were carried out in 444 patients. Overall mortality was 7.8% (n = 35). Thirteen adults with CHD were removed from the final analysis (their surgeries included in our operational costs), leaving the total at 396 studied patients. Organizational expenses for 2015 ($3,460,101) were divided by 444, bringing the cost per patient operated to $ 7,362.00.
Conclusions: The footprint of pediatric cardiac surgery in LMIC is significant. While dimed costly, resources dedicated to global surgery efforts Aggressive healthcare interventions, may result in significant human developmental improvement for LMIC. Global Surgery should be encouraged by international organisms dedicated to global health.

P1070 - DEVELOPMENT OF A SUSTAINABLE NEWBORN CARDIAC SURGICAL PROGRAM IN SIBERIA

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Background: Worldwide, less than 15% of children born with congenital heart disease have access to appropriate medical care. The purpose for this study was to evaluate the effectiveness of Heart to Heart International Children's Medical Alliance's (H2H) multidisciplinary training program to improve the care of

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newborns with congenital heart disease in Tomsk Russia by creating a sustainable center of excellence.
Methods: Retrospective review of cardiopulmonary bypass procedures (CPB) and outcomes in newborns (<30 days of age) from 2005 through 2015. H2H's educational program was between 2006 and 2012. H2H's multidisciplinary educational model incorporates 6 key elements: (1) site assessment including demographics, regional need, medical and societal infrastructure, support for program; (2) development of strategic relationships with key stakeholders; (3) annual medical educational missions with emphasis on education; (4) strategic guidance and programmatic leadership development; (5) bi-directional scholarly exchange and continuing education between annual educational mission trips; (6) facilitation of networking and interaction between H2H sites in Russia.
Results: Table1. Volume of newborns undergoing cardiac surgery increased from 4 (3%) in 2005 to 62 (16%) in 2012 (last year of H2H educational program) and was 37 (9%) in 2015. Newborn mortality decreased from 50% in 2005 to 13% in 2012 and was 11% in 2015. Overall mortality decreased from 9% in 2005 to 4% in 2012 and was 2% in 2015.
Conclusions: The goal of creating a comprehensive, sustainable cardiac surgical program to care for newborns with congenital heart disease can be accomplished. The individualized, multidisciplinary training and educational strategy developed by Heart to Heart International Children's Medical Alliance has proven successful in increasing patient complexity over time with excellent surgical results.

Table 1. Outcomes

	Total Cases	Newborn <30 days	1-12 month	>1 year	STAT 4 (mortality)	STAT 5 (mortality)	Newborn Mortality	Overall Mortality
2005	137	4 (3%)	35 (26%)	98 (71%)	0 (0%)	0 (0%)	2 (50%)	12 (9%)
2006	145	16 (11%)	51 (35%)	78 (54%)	20 (25%)	2 (100%)	5 (31%)	13 (9%)
2007	185	16 (9%)	62 (33%)	107 (58%)	23 (13%)	2 (50%)	5 (31%)	14 (8%)
2008	224	16 (7%)	88 (39%)	120 (54%)	28 (14%)	0 (0%)	2 (12%)	5 (2%)
2009	315	26 (8%)	136 (43%)	153 (49%)	47 (17%)	2 (0%)	4 (15%)	18 (6%)
2010	374	25 (7%)	139 (37%)	210 (56%)	43 (12%)	5 (20%)	4 (16%)	11 (3%)
2011	321	20 (6%)	123 (38%)	178 (56%)	25 (8%)	3 (0%)	2 (10%)	7 (2%)
2012	387	62 (16%)	157 (41%)	168 (43%)	47 (4%)	14 (28%)	8 (13%)	16 (4%)
2013	366	33 (9%)	180 (49%)	153 (42%)	19 (10%)	7 (14%)	4 (12%)	5 (1%)
2014	350	36 (10%)	139 (40%)	175 (50%)	21 (19%)	1 (100%)	3 (8%)	8 (2%)
2015	418	37 (9%)	183 (44%)	198 (47%)	26 (15%)	1 (0%)	4 (11%)	8 (2%)

P1091 - GROWING UP WITH PEDIATRIC CARDIOLOGY

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When I was growing up in Barcelona in the 1950's, pediatric Cardiology did not exist. My father was professor of Pediatrics here in Barcelona and my mother was also a pediatrician. I remember comments such as "Mrs. Rodriguez had a baby, but he died of heart disease". It was accepted because nothing could be done about it. It is remarkable that in only one generation, we have learned, successfully in most cases, to palliate or repair every type of congenital heart disease. In 1961, the 1st Board Examination by the Sub-Board of Pediatric Cardiology was instituted. Surgical repair of most common types of congenital heart disease became commonplace. Mustard and Senning created palliative techniques for transposition of the great vessels. Coronary bypass was invented. Barnard did the first cardiac transplant. Rashkind performed the 1st balloon atrial septostomy. During the 70 s, Jatene introduced the arterial switch procedure for transposition of the great vessels. Prostaglandins were introduced to prevent postnatal ductal closure. Echocardiography revolutionized the diagnosis of infants and children with heart disease. The 1980s saw the development of interventional procedures that transformed the cardiac catheterization laboratory from a diagnostic to a therapeutic tool. The Norwood procedure became an alternative to heart transplantation for hypoplastic left heart syndrome followed by a Glenn and Fontan procedures. Cardiac electrophysiology improved understanding of cardiac arrhythmias resulting in therapeutic ablation techniques for Wolff-Parkinson-White syndrome. In the 1990s balloons were introduced to open structures and devices were developed to close others. MRI and CT scan were used extensively. Fetal echocardiography allowed for the detection of congenital heart disease as early as 16 weeks gestation. Millions of survivors are now living long and productive lives. In 2015, the 1st Board Examination for Adults with Congenital Heart Disease was established.

P1096 - IMPACT AND FUNCTIONING OF SPANISH FAMILIES LIVING WITH A CONGENITAL HEART DISEASE DEVELOPMENT OF A VALIDATED AND CULTURALLY ADAPTED QUESTIONNAIRE

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Congenital Heart Diseases are the most frequent congenital anomalies. They have a significant impact in paediatric morbidity and mortality. CHD represent the congenital disorder with the highest incidence in Spain, with 4,000 new cases every year. Besides, the family's approach may improve or worsen the course of the patient's chronic disease.
Objectives: Know the impact and family functioning of families (including parents, brothers/sisters and the paediatric patient) of a child with complex CHD, who have been subject to more than one surgical/haemodynamic invasive procedure and who will require more interventions in future.
Design: Observational descriptive cross-sectional study project. In this abstract, only the first stage of the study, which aims to develop a validated and culturally adapted questionnaire, is presented.
Subjects and Scope: The sample will include children with heart disease, and their fathers/mothers and brothers/sisters. Data collection will be carried out within families associated to the Foundation Menudos Corazones (1600 associates, with of all ages with CHD).
Method: A questionnaire has been designed using the following validated scales: Family Management Style Framework; "Paediatric Cardiac Quality of Life Inventory; and the Parental Response Style Questionnaire. Two more questionnaires-The Impact of Childhood Chronic Illness on the Family: Psychosocial Adjustment of Siblings, and the Family Social Support Questionnaire have been used after their validation and culturally adaptation to Spanish. The designed questionnaire has been tested with a group of mothers, fathers, children with CHD and brothers/sisters fulfilling the inclusion criteria. The technique used for this pilot stage was the "cognitive interview". The questionnaire will be sent to families associated to the Foundation fulfilling the inclusion criteria for on-line completion.
Conclusions: Obtaining a deep knowledge of families that live with CHD will provide the healthcare team with relevant information for the development of interventions aimed to improve care delivered to these families.

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P1242 - BURDEN OF HEART DISEASES IN CHILDREN ATTENDING CARDIOLOGY CLINIC IN A REGIONAL REFERRAL HOSPITAL IN UGANDA

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Introduction: Patterns and prevalence of heart diseases are different between the high income countries and low income countries. Acquired heart diseases like RHD, cardiomyopathies, pericarditis are still a public health burden while congenital heart diseases still carry a poor outcome among children. The aim of this study was to report the burden and pattern of heart diseases in a semi urban referral hospital in Uganda.
Methods: This was a prospective study in the paediatrics department at a regional referral hospital in Uganda over a period of 20 months. All children from birth up to 12 years who were referred or presented at paediatric department from different areas in the region for an index cardiac evaluation and had their Transthoraxic echo done were included in the study. These children were enrolled and followed up in the cardiology clinic for chronic care. Those who did not turn up for their expected reviews were followed up with phone calls to find out their outcome.
Results: Two hundred and forty children were screened during period and 65.8% had abnormal echocardiological findings. More females than males were had abnormal Echos at 51.9%. VSD were the commonest acyanotic heart disease followed by PDA at 37% and 17% respectively. TOF was highest at 11% followed by Truncus arteriosus at 5.9%. Mean age at diagnosis of congenital heart disease was 28 months (SD 33). RHD was the commonest at 72.5% of the acquired heart diseases. Sixty three percent of children needed and were referred to the national cardiac centre. Only 52% managed to go to the cardiac centre, only 4.4% got surgical intervention. 25% of the children died while 12.7% were lost to follow up.
Conclusion: There is a high burden of heart diseases in our setting with limited access to interventional services.

P1573 - EFFECT AND PROBLEM OF PEER SUPPORT TO THE PARENTS OF FETUS WITH SERIOUS HEART DISEASE

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Introduction: To provide appropriate support to the expectant parents after fetal diagnosis of serious heart disease is essential. However, effective support by medical stuffs to satisfy the client's needs is often difficult because of limited time and manpower. As peer support recently draws attention in the medical or educational fields, we tried to induce it in fetal cardiology.
Methods: Peer support opportunities were arranged to the expectant parents at their second or third visit to fetal cardiac examination. The supporters were selected from the parents of our patients with diseases similar to the client's fetus. At the start of each meeting, the physician briefly introduced the supporter and the clients each other, then they talked for one hour. To prevent the discussion wondering from the subject and to keep time, a nurse attended as a facilitator.
Results: 28 clients were supported by 14 supporters. Two thirds of the fetal heart disease were univentricular heart spectrum for which Fontan-type surgery are expected after birth. The others included biventricular repair candidates and a case of complete AV block. Most important effect of peer support was that the clients could be relieved from anxiety by hearing the supporter's similar experience and coping skills. Also in addition to obtaining useful information such as hospital life, perinatal management and social supports, they could imagine their baby's real future by observing the supporter's children. On the other hand, the outcome of the fetuses is not always good that six fetuses of our series died after birth both pre and post-surgery and that unexpected extracardiac abnormalities including chromosomal abnormality existed.
Conclusions: Peer support is effective for the expectant parents of fetal heart disease. However, we should remember that the supporter's children as well as the client's fetuses are still unstable with their own illness.

P1589 - EFFECTIVE TEACHING METHODS FOR STUDENTS TO LEARN BASIC LIFE SUPPORT AND USE OF AUTOMATED EXTERNAL DEFIBRILLATORS

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Background: In recent years, installations of automated external defibrillators (AEDs) in schools have been increasing in Japan. Despite these efforts, around 40 students die suddenly in schools nationwide each year. To reduce, and even eliminate, occurrence of sudden deaths within schools, students themselves can contribute by acquiring proficiency in cardiopulmonary resuscitation (CPR) methods.
Purpose: We conducted a training session on CPR for students and their families, and examined effective methods of teaching CPR to students.
Method: CPR training sessions consisted of a 40-minute lecture and 60 minutes of hands-on practice. Doctors and nurses who are basic life support instructors authorized by the American Heart Association led the practice portion. Acceptance criteria of practical practice for students were based on those used in an American Heart Association basic life support healthcare provider course. For students and their families, we then conducted a questionnaire on which teaching methods were effective for students.
Results: The number of participating families was 47 (27 students) in the first round and 98 (53 students) in the second round. The questionnaire results showed that in lectures, it was effective to describe CPR in as plain and non-clinical language as possible, and to use animated movies about CPR. In practical skills in these sessions, use of practice manikins and AED trainers used by medical staff were effective means of learning. The instructors judged that students in lower grades of elementary school were capable of using an AED. Students in upper grades of elementary school were capable of performing effective chest compressions and understanding CPR concepts and procedures.
Conclusion: Teaching CPR methods starting at an elementary school age is an effective method for reducing sudden death among students in school.

P1707 - MEDICAL ART THERAPY FOR SYMPTOM RELIEF AND COPING WITH PROCEDURES FOR CHILDREN WITH CONGENITAL HEART DEFECTS UNDERGOING CARDIAC SURGERY

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Background: Creative work can help patients gain insights into life events that may exacerbate their pain and find coping techniques. Nurses spend 90% of their working time with children and may use medical art therapy strategies to offer expressive outlets and pathways to self-organizing for patients.
Purpose: To assess the efficiency of medical art therapy in reducing the anxiety of hospitalized children with CHD undergoing on-pump cardiac surgeries.
Methods: A subgroup of pediatric patients was selected; a single subject research design was initiated with a group of 5 to 8 year-old pediatric patients (n = 40, males – 22, females -18) hospitalized for treatment of CHD. All children with nurses and one pediatric psychologist were proposed to paint, make sculptures and use non-dangerous supplies (disposable gown, gloves, masks, etc.) to express their emotions and fears. The baseline phase began on the first day of each subject's hospital admittance. The intervention phase included art therapy interventions designed to familiarize subjects' with the hospital environment, provide opportunities for control and expression, and respond to subjects' established cognitive structures regarding their medical condition and treatment (at least 5 sessions for each subject). The following assessment instruments were used: the Anxiety Behavior Schedule (ABS), the Children's Health Locus of Control Scale, and the Children's Hope Scale (CHS).
Results: The overall anxiety level according to the ABS was 76.7%. All of the subjects demonstrated reduced externality of locus of control following art therapy intervention. All of the subjects demonstrated high hope on the CHS. The observable anxiety data confirmed reduction in anxiety due to art therapy intervention ($p < 0.05$).
Conclusion: Medical art therapy proved to be effective method in reducing the anxiety among pediatric patients. Opportunities for creative expression can help young patients cope with the psychological challenges of hospitalization and prevent the developmental disorders.

P1865 - ACHIEVING PEDIATRIC CARDIOVASCULAR CENTER EXCELLENCE THROUGH NGO PARTNERSHIP CHILDREN'S HEARTLINK'S SIX YEAR OUTCOMES 2010 TO 2015

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Background: Children's HeartLink, an NGO with almost 50 years of experience, improves access to quality pediatric care. Children's HeartLink's model supports pediatric cardiac programs in low-resource settings to become sustainable regional centers of excellence by working in concert with dedicated medical volunteers to provide training, mentoring and capacity building.
Materials and Methods: In 2008, Children's HeartLink helped found the International Quality Improvement Collaborative for Congenital Heart Surgery in Developing Countries (IQIC) with the mission to reduce mortality and major complications for children undergoing congenital heart surgery. Today, the IQIC has 47 sites in 22 countries with a registry of 61,247 cases. In 2015, open heart surgical data for five Children's HeartLink partner institutions (2,139 cases) were compared to the aggregate of 28 other centers (10,728 cases).
Results: Between 2010 and 2015, Children's HeartLink Partner sites demonstrated overall reduction in postoperative rates of mortality and infection while also increasing surgical case complexity. Unadjusted in-hospital mortality rate decreased from 4.5% to 3.8%, an overall low in 2014 at 3.6%. Unadjusted 30-day mortality rate decreased from 5.2% to 3.8%. Compared to the overall IQIC aggregate, Children's HeartLink partner sites were consistently lower in-hospital and 30-day post-operative mortality; 3.8% vs. 4.9% and 4.0% vs. 5.3%, respectively. Similarly, overall post-operative morbidity reduced within the same time period. Surgical site infections decreased from 7.6% to 1.5%, bacterial sepsis decreased from 20.4% to 3.9%, and other major infections decreased from 25.5% to 5.2%. Meanwhile, case mix demonstrated increased acuity shifting from 74.1% to 64.3% in RACHS-1 Risk Categories 1 & 2 while increasing in higher Categories 3-6 from 25.8% to 33.3%.
Conclusions: NGO support of pediatric cardiovascular centers accelerates surgical outcomes improvement, demonstrated by reduction of post-operative morbidity and mortality following open heart surgery within Children's HeartLink partner institutions, therefore promoting quality in care and center excellence.

P1910 - MOTHER AND INFANT WELLBEING FOLLOWING CARDIAC SURGERY – A FOLLOW UP

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Background/Hypothesis: A secure relationship between mother and infant is vital for optimal child outcomes. Parents' experience of a major medical illness in their infants, including CHD, is extremely stressful and may place mothers' and infants' wellbeing at risk. An earlier study showed a correlation between infant social withdrawal at 2 months and maternal distress following infant cardiac surgery. The infants' CHD ranged from VSD closure to a Norwood procedure. The infants were reviewed at 9 months using the same scales and methods for the follow-up.
Materials and Methods: Infant social withdrawal was identified using the Alarm Distress Baby Scale (ADBB), with >5 as the clinical cut-off score. Maternal distress was assessed using self-report measures for maternal depression (Edinburgh Postnatal Depression Scale (EPDS)), anxiety (Spielberger State-Trait Anxiety Scale (STAI)), and parenting stress (Parenting Stress Index-Short Form (PSI-SF)). Infant social withdrawal in relation to maternal distress was then evaluated.
Results: Twenty-two mother-infant pairs were reviewed. Preliminary data showed maternal distress remained high or even increased for some mothers and resolved for others. A high proportion of infants remained socially withdrawn. Infant withdrawal was not as closely associated with maternal distress at 9 months as it was at 2 months.
Conclusions: Whilst 2-month old infants with CHD subjected to previous cardiac surgery had a substantially increased risk of social withdrawal when their mothers reported experiences of maternal distress, at 9 months preliminary data suggests improvements for some but not all mothers and infants, while others remained at risk. A timely input of services to address mothers' distress following the

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infants' diagnosis of CHD may have seen a more consistent improvement for both mothers and infants at follow-up and may also contribute to better child developmental outcomes.

P1911 - INFANT CARDIAC SURGERY MOTHERS' NARRATIVES

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Background: Congenital heart disease (CHD) requiring cardiac surgery in infancy causes considerable distress for parents.
Materials and Methods: As part of a larger study, 26 mothers of two-month-old infants who had undergone cardiac surgery were interviewed in depth. The mothers were each invited to tell in their own words their experience of the infant's diagnosis, surgery and hospitalisation. They described what had happened to their infant, their experience as mother and what they perceived were their infant's experiences. They were also asked to comment on the interview process. Using qualitative research methodology, a systematic content analysis of the interviews was performed.
Results: Nearly every participant described acute stress symptoms relating to the diagnosis, the infant's surgery and ICU hospitalisation. Yet most reported that the interview with a psychologist with psychotherapy training helped them to think about and integrate what had happened to them and their infant, suggesting that this experience was probably therapeutic.
Conclusions: An appropriately trained mental health professional, providing a safe space for the mother to communicate her story about the diagnosis, her infant's cardiac surgery and hospitalisation, may provide a brief, and very cost-effective therapeutic intervention for these mothers and babies, with only a minimal increase to a surgical programme's costs. The potential to mitigate the trauma and ease maternal distress, may bring about gains for the mother–infant relationship, reducing infant morbidity, and enhancing quality of life for the mothers and infants.

P1939 - NATIONAL PROGRAM OF CONGENITAL HEART DISEASE DESIGN IMPLEMENTATION AND MIDTERM RESULTS

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Background: In order to decrease preventable causes of child mortality, a National Program of Congenital Heart Diseases (NPCHD) was created in 2008. Preliminary studies have shown that in Argentina, yearly approximately 6000 infants are born with congenital heart diseases (CHD), of whom 4300 require surgery. There were nearly 1000 unresolved cases generating waiting lists. This was confirmed with mortality reports between 2002 and 2006. Therefore, a national program was designed. The aim of this study was to describe midterm results based on epidemiologic indicators.
Materials and Methods: A network program was designed, prioritizing timely diagnosis and intervention and consisting of public referral centers, treating centers, and a coordination center. Permanent surveillance was used to improve the program and a unified reporting system was developed.
Results: Different indicators were evaluated comparing the latest results with data from 2012. The waiting list decreased while the number of diagnoses increased. Currently, there are 17500 cases in the Registry; a 47% increase between 2010 and 2015. Diagnosis in infants increased from 40% to 70% and in neonates from 18% to 30% (2010-2015). Age at surgery decreased from 297 days to 160 days (53.8%). In 2015, incidence rate was 5/1000 (range 4-9/1000 according to the different provinces). The number of surgeries increased in every age group, with an increase of 100% in neonates. In 2015, 82% underwent surgery in their area of residence.
Conclusions: The NPCHD has provided a framework to improve access to care and decrease inequalities between provinces. This network was the key to reducing waiting lists and mean age at surgery avoiding morbidity and mortality. Surveillance is essential for the development of strategies. This program is a model of care for other vulnerable populations.

P2066 - THE IMPACT OF TRANSPORTATION ON THE ARRIVAL STATUS AND OUTCOMES IN NEWBORNS WITH HEART DISEASE PERSPECTIVE FROM A LOW AND MIDDLE INCOME COUNTRY

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Background: With increasing availability of newborn heart surgeries in low and middle-income countries (LMICs), health systems challenges, specifically relating to transport may need to be addressed.
Objectives: To identify determinants of clinical status at arrival of neonates with congenital heart disease (CHD) transported to a pediatric heart centre, and to study the impact of the arrival status on clinical outcomes.
Methods and Results: 100 consecutive neonates with CHD (January 2016- December 2016) transported to a pediatric cardiac program in Southern India were studied prospectively using a structured database that captured relevant demographic, clinical and laboratory information and specific transport variables. Physicians were available in 34, nurses in 53 and neither in 13 transports; 35 babies were ventilated; 14 required ventilation <2 hours after arrival. Of 43 babies with duct dependent circulation, 27 (63%) were transported without prostaglandin infusion. Problems identified on arrival included hypothermia (23), hypoglycemia (9), cardiac arrest (4), endotracheal tube block (1), high FiO2 in shunt lesions (10), end organ (liver/kidney/cerebral) injury (26) and blood stream sepsis (20). Following stabilisation, 65 were offered cardiac surgery (59 underwent surgery) and 16 underwent catheter intervention, 9 parents opted for comfort care (3 with severe end organ damage), 10 required medical management. Three of 5 deaths before surgery resulted from poor arrival status. End organ injury at arrival was seen in 4/5 patients who died (all 3 surgical deaths and 1 of 2 deaths after catheter intervention). End-organ injury was strongly associated with all cause mortality (27% vs. 4%, $p < 0.003$)
Conclusion: In the absence of organised transport systems, a quarter of neonates with CHD arrived with end organ injury and this significantly impacted their mortality and morbidity. Robust transport systems need to be integrated in plans to develop newborn and infant heart surgery in LMICs.

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P2166 - TRANSCATHETER CLOSURE OF PATENT DUCTUS ARTERIOSUS WITH THE AMPLATZER DUCT OCCLUDER IN SUB SAHARAN AFRICA

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Objectives: To analyze the safety and efficacy of PDA trans-catheter closure in Shisong Hospital Cameroon.
Background: During an 8 year period, a Cardiac Centre was designed and developed in Sub-Saharan Africa. Three partners worked towards the realization of this project (Associazione Cuore Fratello, Associazione Bambini Cardiopatici nel Mondo, Tertiary Systers of St Francis).
Methods: From 2010 to 2015, 45 patients with clinically significant PDA (30 and 15 adults) were treated. Median age was 11yrs (range 2-33years) while median weight was 33 kg (range 14-58 kg). Indications for closure were: cardiac failure (n = 4), failure to thrive (n = 2), left ventricular overload (n = 43), arrhythmias (n = 2). The duct was defined as large (pulmonary pressure > 3/4 systemic pressure), moderate (pulmonary pressure between (1/4-3/4) of the systemic pressure and (QP/QS > 1.5), restrictive (normal pulmonary pressure and QP/QS = 1). Procedures were performed under general anesthesia. The Amplatzer Duct Occluder (ADO I) and MVSD devices were used.
Results: The duct was restrictive in 4 pts, moderate in 2 pts and large in 1 pys. The mean PDA diameter was 6 mm (range 2.5-16mm). Successful device implantation was performed in 37 out of 45 (82%). Three subjects were not closed because of Eisenmenger syndrome. Complications occurred in 2 subjects (8%) who had embolization: in one subject surgery was needed while in one the device was retrieved and the PDA successfully closed using a MVSD device. MVSD was used in three patients while ADO I was used in the other subjects.
Conclusion: Successful and safe percutaneous treatment of PDA can be performed in Sub-Saharan Africa.

P2264 - DEVELOPMENT OF AN AUSTRALIAN NATIONAL POLICY FRAMEWORK FOR PAEDIATRIC CARDIAC AID WORK

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Background: Healthcare systems and services are receiving increasing numbers of aid-related referrals for cardiac surgical intervention. Paediatric cardiac interventions and surgery are high-cost, resource-intensive services, in demand for both non-funded and privately funded overseas patients. Current practice leads to competition and an individualistic approach to the provision of medical care and raises many ethical and practical issues.
Methods: Paediatric clinical ethics services in Australia collaborated in discussions to develop a potential policy framework for on-shore paediatric cardiac medical aid work. We used a narrative-medicine based approach to explore the stories of this patient group and the medical teams that work with them. We collected qualitative data from families regarding their experiences and their advice for improvements, and from their healthcare teams including social workers. Quantitative data were collected regarding the resourcing costs for different surgeries and the methods of funding from the administration departments of hospital networks and aid organisations involved in the processes.
Results: The completed draft policy framework will be presented to the combined cardiac and cardiac surgical departments in public paediatric health care services in Australia for further discussion and ratification. The policy aims to reduce practice variability, improve outcomes and experiences for patients and families who receive care within the Australian Health-care system and improve distributive justice and rationalisation of limited public resources.
Conclusion: We will present a discussion based on the ethics and practicalities of medical aid work and medical tourism for paediatric cardiac services in an Australian context. The draft framework will be used as the basis for the wider discussion of the provision of services and the ethical practicalities in different nations and health-care systems in the Asia-Pacific region.

P2287 - THE NAMIBIAN CHILDREN'S HEART PROJECT A SIX YEAR REVIEW OF MISSION PATIENTS AND OUTCOMES

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Background: Before 2007 there were no cardiac services in Namibia. A service was initiated in 2009 and heart surgery from 2010. However, surgery cannot be offered to infants or patients with complex disease. Through a public-private partnership patients are referred to the Christian Barnard Memorial Hospital in South Africa.
Materials and Methods: This is a retrospective review of the 200 patients referred between 2009 and 2015. Demographics, diagnosis, interventions, morbidity, mortality and follow-up were reviewed. Data was entered using RedCap. Continuous variables expressed as means or medians and categorical variables as percentages. Linear regression models compare variables. Cox regression and Kaplan- Meier curves assess survival.
Results: 188 (94%) had congenital and 12 (6%) acquired disease. Age at presentation ranges from 1 week to 33 years. Lesions include right and left ventricular outflow obstruction, atrio-ventricular and ventriculo-arterial discordance, anomalies of systemic and pulmonary venous drainage, single ventricle, left to right shunts, Rheumatic Heart Disease and Takayasu arteritis. Twenty five (13%) had percutaneous interventions. The majority had definitive repair whilst 30 (15%) had palliative procedures. Palliation was either staged or definitive. RACHS categories were 2 to 5. Early (<30) and late (>30 days) postoperative mortality was 6 (3%) and 2 (1%), respectively. A quarter defaulted follow-up.
Conclusion: Patients present late with consequent added complexity. The relative paucity of newborns with critical CHD is revealing. The number of patients defaulting follow-up represents health system challenges in a developing country. A comprehensive, sustainable local surgical service remains our goal, however in the medium term this humanitarian programme with a unique funding model has achieved good outcomes for indigent patients.

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P2311 - EFFICACY OF A CLINICAL TOOL TO DETECT CONGENITAL HEART DEFECTS IN A RESOURCE LIMITED COUNTRY

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Background: Neonatal screening programmes have incorporated pulse oximetry for early diagnosis of CHDs and have been cleared by the FDA for use in newborns. Detection rates for CHD by prenatal ultrasound is ~25-50% and by postnatal newborn physical exams is ~25-50%. The immunization clinic provides a unique opportunity to screen missed and undiagnosed cases especially from non institutional deliveries. This can serve as a second checkpoint beside neonatal screening for the detection of CHD. The present study is undertaken to develop a cost effective screening tool (score) for early detection of CHD and further investigate that how effective is the clinical screening score in detecting undiagnosed or missed cases of congenital heart disease?
Materials and Methods: Children presenting in the OPD at 6weeks of age for immunization or any child presenting for first time (outborn delivery) till 6 months of age in the immunization clinic were subjected to detailed history and examination and findings were recorded on a predesigned proforma and CHD score was recorded. These children were taken for echocardiography for confirmation of the diagnosis of CHD. The sensitivity and specificity of the test was calculated at different cut-off scores.
Results: It is continuing project with a target of screening at least 500 newborns in next 6 months, as a pilot programme initial 30 infants were screened, out of which 4 had CHD. The remaining newborns were normal and had a score of less than 3. All 4 cases of CHD had a score of more than 3 with severity increasing with score.
Conclusions: History and examination based tool can be utilised as an effective method of second screening of newborn in the immunisation clinic for early identification of CHDs.

P2440 - TELL ME ONCE TELL ME SOON PARENTS' PREFERENCES FOR GENETIC CONSULTATIONS FOR CONGENITAL HEART DISEASE

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Background and Hypotheses: An important aspect of designing healthcare services is to understand how individuals choose between services. Discrete choice experiments (DCE) are increasingly used in health services research to understand such choices. In response to the changing landscape of cardiac genetics, we developed a DCE to identify parents' preferences for genetics services for families affected by congenital heart disease (CHD).
Materials and Methods: One-hundred fourteen parents of a child diagnosed with CHD requiring surgery were recruited via the Sydney Children's Hospital Cardiac Service (participation rate: 54%), of whom 22% had previously accessed genetics services for CHD. DCE attributes (n = 4) and levels (n = 3) were developed in consultation with experienced clinicians, and combined in an orthogonal design of nine choice sets. Each participant received all nine choice sets and indicated their preferred consultation type, and whether they would accept a referral to that consultation if offered. Mean choice coefficients were analysed using both conditional logit and mixed logit analyses to incorporate the extent to which there was heterogeneity across participants in what influenced their choices.
Results: Participants overwhelmingly preferred genetics services for CHD that involve only one consultation with both a clinical geneticist and genetic counsellor, that occur with a minimum waiting period of two weeks, and where information is provided verbally and via a website. Such a service, if offered, would result in participation by 93% of parents in our study.
Conclusions: Incorporating a genetic counsellor in all CHD-related genetics consultations and reducing waiting time would result in high uptake by parents. Independent of parent preference, there are clear advantages to such an approach. As we have shown elsewhere, CHD in a child can have profound psychological consequences for families and access to genetic counselling can provide a valuable component of the genetics consultation.

P2443 - THE NEED FOR UNIVERSAL SCREENING OF HEALTH RELATED QUALITY OF LIFE IN YOUNG CHILDREN WITH COMPLEX CONGENITAL HEART DISEASE AND THEIR FAMILIES

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Background and Hypotheses: Studies examining health-related quality of life (HRQOL) in young children with congenital heart disease (CHD) and their families are sparse. This study aimed to assess maternal perceptions of HRQOL in two groups of young children with CHD; those following a single ventricle (SV) surgical pathway and those requiring biventricular repair. It also aimed to identify the demographic, clinical, psychological and social determinants of HRQOL in these children and their mothers. It was hypothesised that children with SV anomalies and their mothers would have poorer HRQOL than the biventricular group.
Materials and Methods: Participants were recruited via the Sydney Children's Hospitals Network Cardiac Service. Mothers of a child aged 1-5 years with either SVCHD (pre-Fontan) or CHD requiring neonatal biventricular repair and cardiopulmonary bypass were invited to participate. Consenting mothers completed a validated survey instrument assessing HRQOL and a range of demographic, clinical, psychological and social factors.
Results: Eighty-six mothers participated (response rate: 48%). Overall, children with CHD had lower parent-reported HRQOL than normative data and 58% of children scored below the cut-off indicative of a need for clinical investigation. Poorer child HRQOL was strongly associated with SV anomalies (Beta = -0.43, $p < 0.0001$), physical comorbidity (Beta = -0.26, $p = 0.004$), feeding problems (Beta = -0.27, $p = 0.003$), and greater maternal psychological stress (Beta = -0.18, $p = 0.04$). Together these variables accounted for 52% of the variance in child HRQOL. Poorer maternal HRQOL was strongly associated with poorer family functioning (Beta = 0.62, $p < 0.0001$), greater maternal psychological stress (Beta = -0.23, $p = 0.005$), and 'difficult' child temperament (Beta = -0.19, $p = 0.006$); accounting for 73% of the variance in maternal HRQOL.

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Conclusions: Biopsychosocial factors accounted for over half the variance in child HRQOL, and over 70% of the variance in maternal HRQOL. Universal screening of HRQOL in children with CHD and their parents, coupled with early psychological intervention for those at risk, could improve HRQOL in this population.

P2445 - QUALITY OF LIFE EMOTIONAL WELLBEING AND BEHAVIOURAL ADJUSTMENT AS EXPERIENCED BY YOUNG CHILDREN WITH CONGENITAL HEART DISEASE A SYSTEMATIC REVIEW

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Background and Hypotheses: Current understanding of quality of life (QOL) in young children with congenital heart disease (CHD) is limited. Better understanding of QOL during these formative years could enable earlier identification of difficulties and more timely intervention. This systematic review aimed to synthesise and critically appraise the literature examining QOL, emotional wellbeing and behavioural adjustment in children aged ≤5 years with CHD. It also aimed to identify demographic, clinical, psychological and social determinants of these outcomes.
Materials and Methods: A systematic literature search was performed using five electronic databases; Medline, Embase, PsycINFO, CINAHL, Scopus. Articles were included if they reported on parent-proxy or self-reported outcomes associated with QOL or emotional, social or behavioural functioning for children with CHD with a mean age ≤5 years. Risk of bias was appraised for each article using the QualSyst framework.
Results: Thirty-eight articles were identified for review (31 cross-sectional studies, 3 prospective case-cohort studies, 2 prospective cohort studies, 2 randomised controlled trials), with data from 3,841 children with CHD. Most studies assessing QOL (6/8 studies) found poorer QOL in children with CHD compared to normative data or healthy controls. Behavioural and emotional functioning was poorer in children with CHD than comparator groups in 13/20 studies. All studies assessing adaptive behaviour and functional living skills (11/11 studies) reported impairments, with scores for 8-79% of children indicating a need for clinical intervention. Parental stress was the most common predictor of QOL-related outcomes.
Conclusions: Young children with CHD demonstrate greater impairment than comparator groups in QOL, emotional well-being and behavioural functioning, highlighting the need to identify and respond to these difficulties in clinical settings. The prevalence and determinants of these impairments vary markedly and research effort should be directed towards developing a consistent approach to assessment and more accurately characterising these outcomes in this population.

P2461 - A PEDIATRIC CARDIOLOGY TELEMEDICINE NETWORK IN NORTHEAST BRAZIL MAIN RESULTS FROM FIVE YEARS OF EXISTENCE

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Background: Providing appropriate health care for children with congenital heart disease in middle and low-income countries is a constant challenge. This situation becomes more critical when the country has a continental size, like Brazil, because the few pediatric cardiologists concentrate in large urban centers leaving vast areas completely unconvered. Until 2011, this was the reality in Paraíba, a state from northeast Brazil.
Material and Methods: A partnership between the Government of Paraíba and the NGO - Heart Circle was established in October/2011. The work method was the establishment of a Network of services, with the aid of telemedicine, to provide an integral line of care from screening to surgery and post-operative care.
Results: From 2012 to 2016, 146.422 babies were screened for heart defects and 1.951 abnormalities were identified. The screening program covered 65.5% of the target population (neonates with 34 or more weeks of gestational age in the participant centres) and 49.1% of all births in the public health system in the State in 2012; and in 2016 this coverage reached 95.7% of the target population and 95.1% of all births. Detection of congenital heart disease increased from 4.09 to 13.2 per 1000 live births ($P < 0.001$). Over 20.000 consultations and echoes were supervised via telemedicine and a total of 532 cardiac operations were performed. Over 5.000 professionals received live and/or online training.
Conclusion: In five years, the pediatric cardiology network established a universal screening program for CHD in Paraíba and structured the care for children at different levels according to specific needs. We believe that this work-model can be adapted to other countries and realities around the world.

P2471 - CHD IN THE TIME OF HASHTAGS AND TWEETS E HEALTH LITERACY AND PREFERENCES FOR E HEALTH RESOURCES IN PARENTS OF CHILDREN WITH COMPLEX CONGENITAL HEART DISEASE

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Background and Hypotheses: The rapid increase in Internet-based healthcare information, or eHealth, has not been matched with an increase in research examining how patients and families use and experience this information, and any associated clinical implications. This is particularly the case in paediatric cardiology, where there is little existing research. This study aimed to: (a) examine eHealth literacy, beliefs and behaviours in parents of children with complex congenital heart disease (CHD), and (b) identify parents'

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preferences for the content, format, features and functions of CHD-related eHealth resources.
Materials and Methods: Families of a child born between 2008-2011 and diagnosed with CHD requiring surgery at the Heart Centre for Children in Sydney, were mailed a survey assessing eHealth literacy, beliefs and behaviours, and preferences for CHD-related eHealth resources.
Results: One-hundred thirty-two parents (83 mothers, 49 fathers) completed the survey (response rate: 50%). Mothers (96%) were more likely to access eHealth resources than fathers (83%, $p = 0.009$). Despite high eHealth use, eHealth literacy was relatively low, with widespread gaps in parents' awareness of, access to and communication about eHealth resources. Over 50% of parents reported that decisions regarding their child's healthcare were influenced, to some extent, by eHealth. Barriers to doctor-patient communication about eHealth included limited consultation time and concern about doctor's disapproval. Participants demonstrated a strong desire for 'eHealth prescriptions' (or recommendations) from their healthcare team, and perceived a range of eHealth topics as highly important, including treatment-related complications, and physical, cognitive and emotional development in children with CHD.
Conclusions: This is the first time eHealth literacy has been assessed in parents of children with CHD. Results demonstrate a need for greater education and support as families navigate the complex landscape of CHD-related eHealth. 'eHealth prescriptions' from the cardiac team are highly desired by families and may be a good starting point.

P2474 - BIOPSYCHOSOCIAL CONTRIBUTIONS TO RISK AND RESILIENCE IN INFANTS WITH COMPLEX CONGENITAL HEART DISEASE AND THEIR PARENTS

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Background and Hypotheses: Early life experiences shape brain development, the immune system and responses to stress. For infants with complex congenital heart disease (CHD), experiences associated with diagnosis and treatment can have enduring developmental consequences. This prospective case-control study examines the prevalence and predictors of psychological morbidity in parents following CHD diagnosis. It also examines how biopsychosocial factors may influence developmental outcomes in infants with complex CHD.
Materials and Methods: Parents of babies with a fetal or postnatal CHD diagnosis requiring surgery in the first six months of life ($n = 193$) or healthy fetal morphology scan at 18-20 weeks gestation ($n = 120$) complete neuropsychiatric interviews (at 3-months post-diagnosis and 12-months postpartum) and psychological measures (3-months post-diagnosis and 3-, 6-, 12-months postpartum). Salivary cortisol is collected as a biological indicator of stress reactivity in mothers at 32-weeks gestation, and in mothers and infants at 12-months postpartum. Mother-infant interaction is assessed at 6-months (CARE-Index), infant developmental outcomes are assessed at 12-months (Bayley Scales of Infant Development, Strange Situation), and clinical data are collected via medical records.
Results: Three months post-diagnosis, 45% and 43% of mothers (57% and 47% of fathers) in the fetal and postnatal CHD groups respectively, reported anxiety warranting clinical intervention compared to 19% of mothers (12% of fathers) of typically-developing babies. After controlling for education and income, maternal antenatal anxiety after fetal diagnosis was strongly associated with self-blame ($Beta = 0.28$, $p = 0.02$) and disengagement ($Beta = 0.26$, $p = 0.03$). At 6-months, >60% of mothers in the postnatal CHD group perceived their infant as 'difficult' compared to 8% and 0% in the fetal CHD and control groups, respectively. Biopsychosocial contributions to infant developmental outcomes are being investigated.
Conclusions: Infants with complex CHD experience early adversity and their parents report marked psychological stress. We have developed and are currently evaluating an integrated health system response to these needs.

P2496 - ACTIVE SEARCH FOR CONGENITAL HEART DISEASE MAIN RESULTS FROM FOUR YEARS OF MEDICAL MISSIONS IN NORTHEAST BRAZIL

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Background: Congenital Heart Disease (CHD) is a major public health problem and the main cause of mortality in neonates when infectious diseases are excluded. In developing regions, the lack of CHD diagnosis can lead to clinical worsening or even death. In such scenario, an annually medical mission performs an active search for CHD in northeast Brazil.
Material and Methods: This is a retrospective study based on the data from an active search (called "The Heart Caravan") performed between 2013 to 2016. The active search covers 13 cities (only 8 in the first year) and takes place in two stages. In the first, local health centres perform a screening of children with symptoms or history of cardiovascular diseases. In the second phase, a multidisciplinary team (paediatric cardiologists, nutritionists, psychologists, nurses, social service and volunteers) performed health consultations and echocardiograms in children and pregnant women to diagnosis CHD or monitored previously diagnosed patients. A frequency analysis was performed.
Results: A total of 4.464 patients were seen in the Caravans. In 2016, however, there were 616 patients seen in relation to the microcephaly outbreak. From the 3.848 patients seen by the cardiology teams, there were 3.532 children and 316 foetuses. In 2016, there were approximately 60% of new patients. The main reason to participate in the caravan was a known CHD (21.11%) or the presence of a heart murmur in children (19.8%). Most patients had a poor background (more than 50% using government social benefits). A total of 1.092 CHD (126 complex CHD) were diagnosed or monitored during the Caravans. In relation to

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pregnant women, only 30 (9.6%) showed an abnormal foetal echocardiogram.
Conclusion: The Heart Caravans identified and monitored an expressive number of CHD in childhood. These patients were inserted in the health services for appropriate management.

P2503 - LIMITED RESOURCES LONG TERM CLINICAL OUTCOMES IN A PEDIATRIC CARDIAC SURGICAL PROGRAM IN NIGERIA

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Background/Hypothesis: Congenital heart disease (CHD) is a leading cause of birth defect-associated infant illness and death. The incidence of CHD is 0.8% and virtually all developed countries have sustainable surgical programs. The ability to establish programs in resource limited environments is critical as nearly half (48%) of the deaths due to CHDs occur during infancy. The aim of this study was to assess a newly established sustainable pediatric cardiac surgical program in Nigeria with regard to number of surgeries and cardiac catheterization procedures performed annually. Secondary aims were to identify common comorbidities, average length of stay, and mortality.
Methods: A prospective study design was utilized to follow a cohort of 48 pediatric patients who received cardiac surgery from January 2015 – January 2017. 349 patients were screened from a list of 1566 referred patients. Each patient admitted to the hospital was reviewed by a physician and data was recorded and retrieved from medical records. This study included all patients that had surgical intervention for CHD. Variables collected for data analysis were number of surgeries and cardiac catheterization procedures, number and type of comorbidity, average length of stay, and mortality. Descriptive statistics was used to analyze the data.
Result: Upon establishment of the pediatric cardiac surgical program 48 cardiac surgeries and one cardiac catheterization procedure was successfully performed in 2 years (25 surgeries in year one and 23 surgeries in year two). Major comorbidities were infection, reintubation and bleeding. Findings revealed that average length of stay was 10 days, while mortality was 5 patients.
Conclusion: The establishment of a cardiac surgical program in a resource limited environment is possible. Limitations in completing more surgeries include cultural differences, limited space, nurse and support staff education. Establishing sustainable educational programs in underserved environments can lead to the provision of care for thousands with CHD.

P2512 - ESTABLISHING A PEDIATRIC CARDIAC SURGICAL PROGRAM IN A RESOURCE LIMITED ENVIRONMENT THE NIGERIAN EXPERIENCE

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Background/Hypothesis: Congenital heart disease (CHD) is a leading cause of birth defect-associated infant illness and death. Incidence is 0.8% and virtually all developed countries have sustainable programs for care. The ability to establish programs in resource limited environments is critical as nearly half (48%) of the deaths due to CHD occur during infancy. The aim of this study was to describe the establishment of a sustainable pediatric cardiac program with multidisciplinary support in a resource limited country and to discuss limitations.
Methods: This descriptive study examined the process of establishing a surgical program over a two year period. A work group was convened with members from Hospitals for Humanity (HFH) a NGO, Garki and National Hospital, Abuja, Nigeria. Current available resources were identified including surgical centers. Agreements were then established with governmental programs. Lastly, HFH connected with medical sub-specialists across the world interested in building a program vs. planning for a single medical mission trip.
Results: Based on published and unpublished data, the work group made recommendations for a standardized approach to educational program development. The organization HFH served as a fundraising instrument to pay for care for patients and acquire equipment for the PICU and operating room. The staff included: surgeons, cardiologists, PICU physicians, PICU nurses and support personnel. The program allowed for an operating room, catheterization lab, 4 PICU and 10 in-patients beds. A total of 48 cardiac surgeries were performed. Although plans are underway to continue the program, limitations included corruption, resources and exposure.
Conclusion: Key factors for success were local physician and government involvement, committed medical staff, and publicity. An unforeseen factor was having ex-patriots return to their country of origin to provide communication and education. A consistent and long term approach is suggested, lengthening the time of each medical mission until a sustainable program is established.

P2548 - PEDIATRIC CARDIOLOGY EXCHANGE PROGRAM BETWEEN GONDAR UNIVERSITY MEDICAL CENTER GONDAR ETHIOPIA AND SHAARE ZEDEK MEDICAL CENTER JERUSALEM ISRAEL

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We describe an ongoing collaboration between the Medical Center at Gondar University, Ethiopia, the Shaare Zedek Medical Center in Jerusalem, Israel and the American Jewish Joint Distribution Committee (JDC). As part of a more general collaboration between our three organizations, starting in March 2012, the division of pediatric cardiology in Jerusalem has partnered with the department of pediatrics in Gondar with reciprocal doctor exchanges facilitated by the JDC. The JDC supplied robust cardiology stethoscopes, electrocardiogram and portable echocardiogram equipment for the Gondar pediatrics department. One author (DLF) has traveled to Gondar for several two week periods to teach the pediatricians cardiac physical examination including auscultation with up to 28 digital demonstration headphone stations, electrocardiography and echocardiography and one author (ZT) has traveled for several three month periods to

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Jerusalem to work integrally with the pediatric cardiology division. We performed a small pilot study screening school children in the Gondar region for asymptomatic clinically evident but undiagnosed rheumatic heart disease finding a rate of 2%. We have also developed a program where ECG and echocardiogram loops are sent by electronic mail for long distance consultation. We are excited with this grass roots model advancing the process of integrating advanced pediatric cardiac care into the local infrastructure in an underdeveloped country.

P2566 - SHINE A SAVE HEART INITIATIVE PROVIDING FREE CURATIVE TRANSCATHETER TREATMENT FOR CHILDREN WITH CARDIAC SHUNT LESIONS IN SOUTH INDIA

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Background: Cardiac shunt lesions form a significant proportion of the disease burden amongst children born with congenital heart disease. While Atrial septal defects (ASD), Ventricular septal defects (VSD) and Patent Ductus Arteriosus (PDA) constitute the major bulk of this group, other shunt lesions such as Coronary fistula, Ruptured sinus of valsalva (RSOV), Aortopulmonary window present from time to time. Majority of these shunt lesions are amenable for transcatheter closure and when undertaken appropriately, offers complete cure at a significantly lower cost than surgery.
Materials & Methods: We report our institutional experience of 410 patients with 395 shunt lesions addressed by the transcatheter route, between April 2014 and January 2017. 21 procedures were self-funded by the patients and 374 procedures were done free of cost for economically challenged children. Of the 374 procedures, 172 (45.8%) were ASD device closure, 115 (30.6%) VSD device closure, 87 (23.2%) PDA device closure, 1 (0.2%) Coronary fistula embolisation and 1 (0.2%) RSOV occlusion. Funding was by an amalgamation of state sponsored welfare schemes, donations from Non Governmental Organisations (NGO), subsidies by the Medical device supplier and by our institution under the project SHINE (Save Heart Initiative). State sponsorship was 50% of the total expenditure and the rest was from donors chiefly the US based NGO SAMAHOPE and the India based NGO Have a Heart Foundation (HAHF).
Results: Four patients of the 172 (2.3%) who underwent ASD device closure had immediate device migration necessitating surgical retrieval along with ASD patch closure. There were no other morbidities or mortalities.
Conclusion: In developing countries where cardiac care is not entirely state sponsored, it is feasible to provide free care safely through a bouquet of charitable measures. Transcatheter management offers a significant reduction in cost burden facilitating the spread of funds amongst more deserving children.

P2612 - LOW AND MIDDLE INCOME COUNTRY PERSPECTIVE OF CONGENITAL HEART DISEASE ASSOCIATED WITH DOWN SYNDROME – PREVALENCE SPECTRUM TREATMENT OUTCOMES AND BIASES

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Background: There is little published data from Low and Middle-Income countries (LMICs) on Down Syndrome (DS)-associated-CHD or their treatment outcomes. Social stigma, and ignorance often result in delayed diagnosis of CHD and denial of definitive treatment in resource-limited environments.
Objectives: To determine prevalence and spectrum of CHD, and in-hospital outcomes of cardiac surgery/intervention among patients with DS presenting to Pediatric sub-specialty clinics of a tertiary-care center in South-India.
Methods and Results: Clinical records of 764 consecutive patients with karyotype-proven DS who presented (November 2005–January 2017) at a tertiary hospital with well-developed pediatric genetics and cardiology services were analyzed. Of these, 57% primarily presented to pediatric genetics; 31% to pediatric cardiology services and12% presented to other pediatric specialties. Echocardiography was done irrespective of cardiac symptoms/signs in 96%. CHD was present in 44% among those evaluated in genetics, 92% of those who primarily presented to cardiology and 62% of those who presented to other subspecialties. Overall prevalence of CHD was 62%. Most common lesions were complete -atrioventricular-canal-defect (24%), simple-VSD (22%), ASD (19%), PDA (15%), TOF (4%). Surgery/intervention was offered to 256 patients and carried out in 158 (62%). In 38% (n = 98), caregivers refused surgery/intervention. Heart surgery included VSD closure (57), AV Canal repair (44), transcatheter treatment (PDA occlusion-22, ASD closure-3). Outcomes: In-hospital mortality was 2.5%. Morbidity included prolonged (>4 days) ICU stay (24.6%), prolonged (>48 hours) ventilation (12.6%), respiratory complications (10.8%), bloodstream-infection (3.8%).
Conclusions: Echocardiographic screening of all patients with DS is recommended for early diagnosis of CHD. Majority of DS-associated CHD can be treated with low mortality. Prolonged ICU-stay, ventilation and respiratory complications contribute to morbidity must be anticipated and managed appropriately. Biases still lead to denial of treatment to large number of DS patients with CHD in LMICs even in high-expertise centers.

P2703 - IMPLEMENTATION OF NEWBORN SCREENING FOR CHD IN RURAL CHINA

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The concept of newborn screening for congenital heart disease (CHD) developed slowly from small studies between 2002 and 2009 to larger population based studies in Europe and the U.S. These studies and associated advocacy provided key data driving the US Department of Health and Human Services to formally add CCHD screening to the Recommended Uniform Screening Panel (RUSP) in the United States in 2011. Today, more than 30 international countries have either implemented or are in the process of evaluating newborn screening using pulse oximetry. Challenges remain however, particularly in low-resource health settings. The presentation will focus on implementation methods and results from screening more than 20,000 well-appearing newborns across 18 birth facilities in rural Sichuan Province China, utilizing a low-cost, mobile-phone pulse oximeter to measure blood-oxygen saturations at two points – 12 and 48 hours after birth. Overall incidence of congenital heart disease (CHD) in the region is higher than the national average at 11.74% (vs 6.75%), with 20%~30% of CHD patients dying within the first year.60 babies failed the screening based on a predetermined algorithm. Of those, 18 had a resulting diagnosis of Critical Congenital Heart Disease and additional 25 babies had a secondary

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diagnosis. Notably, more than 50% of failed screens resulted in a diagnosis of previously unrecognized neonatal pneumonia. Neonatal infection (sepsis and pneumonia) rank among the most common causes of newborn mortality, accounting for more than a quarter of newborn deaths each year. The sensitivity and specificity of screening in identifying non-target conditions combined with reducing missed or delayed diagnosis of congenital heart diseases make this an increasingly high impact screen. The project deployed a train-the-trainer implementation model supported by public health and clinical leadership and robust data collection to identify and improve gaps in follow-up diagnostics, referral pathways and treatment infrastructure.

Figure 1.

Figure 2.

Figure 3.

P2717 - OPTIMIZING OUTCOMES THROUGH GLOBAL COLLABORATION

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Background: Access to care for children with congenital heart disease in developing countries is often inadequate, and limited resources pose additional challenges to centers that provide pediatric cardiac surgery. Congenital heart surgery carries considerable risk for death, especially in resource-limited settings. Following the 2008 Global Forum on Humanitarian Medicine in Cardiology and Cardiac Surgery meeting in Geneva, the International Quality Improvement Collaborative (IQIC) was established as a solution. Utilizing quality improvement strategies, three key drivers were identified to provide targeted education via monthly webinars in an effort to reduce mortality associated with congenital heart surgery. The key drivers are safe perioperative practice, infection reduction and team based practice through nurse empowerment.
Materials/Methods: Webinars related to each key driver were created by subject matter experts. Slides were disseminated prior to live presentations for individual sites to make accommodations for translation. Following each live presentation, time was allotted for interactive questions, answers and discussion. Each presentation was recorded and uploaded to the IQIC site for participants to copy, and use at their discretion. Follow up questions were emailed through the IQIC and responses were provided by presenters. Based on participant feedback and needs assessment survey data, advanced nursing modules were created and presented to meet ongoing and evolving educational needs.
Results: Since the programs' inception in 2010, 92 webinars have been created and presented. Multiple sites from around the globe were present for each live webinar. Discussion following each presentation included a rich exchange of clarifying ideas and sharing unique insights to programmatic challenges.
Conclusions: The IQIC has proven to be successful by providing interactive and interdisciplinary education, and collegial support in the subspecialty of pediatric cardiac critical care. Longstanding relationships have been forged in all disciplines.

P2723 - EXPERIENCE WITH LATE CORRECTION OF TETRALOGY OF FALLOT IN THE SETTING OF A HUMANITARIAN COLLABORATION PROGRAM

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Objective: To report our experience and results of late surgical correction of patients with tetralogy of Fallot (TOF) in the setting of a humanitarian collaboration program.
Patients & Methods: Retrospective analysis of the perioperative course of all patients undergoing correction of TOF older than 2 years of age at the University Hospital Bern or the University Children's Hospital Rabat between November 2011 to November 2016.
Results: 25 children (mean age: 68.9 months, range:24-156 months; 40% female) underwent total correction of TOF.

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Two patients were initially palliated with a shunt with subsequent correction after 108 and 24 months. Preoperative oxygen saturation was 85 + /- 11%; mean hemoglobin was 146 + /-23 g/l. Preoperative Mean RV/PA gradient was 84 + /-21 mmHg with a Nakata-Index of 162.6 + /-55.6mm2/m2. A valve-sparing correction was performed in all patients. Ventilation time 28.7 + /- 14.6 hours. Length of stay was 11.7 + /-3.3 days. Mortality was 0%. No stroke occurred. No permanent pacemaker had to be implanted. All patients were examined by echocardiography at 1months postoperatively. Due to the character of the program, follow-up is 60% complete with a mean of 17 months (range: 3-43mo). Last echocardiography demonstrated less to mild pulmonary stenosis in 16patients, moderate in 9patients, no severe pulmonary stenosis. 2 patient underwent reoperation (1 patch dehiscence, 1 recurrent pulmonary stenosis).
Conclusions: Late correction of TOF can be safely performed in older children with good postoperative results and low morbidity comparable to reported results for "timely" correction in infants. A valve-sparing correction might be possible in the majority of those patients. Follow-up study for the evaluation of the development of right ventricular and pulmonary valve dysfunction is needed and in progress.

P2729 - PEDIATRIC CARDIAC DEVELOPMENT ASSISTANCE IN IRAQ; 2010 2016

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Background: Pediatric cardiac surgery in Iraq prior to 2010 was available in only 2 cities, Erbil and Baghdad. We were asked to assist in the development of pediatric cardiac services in 2009 and began in 2010. Four sites were identified, in 2010 Sulimaniyah and Nasiriyah; Najaf and Basra were added in 2012. We present the results of this unique country-wide program.
Methods: We queried our database for; number of trips, patient operations, RACHS classifications, 30-day/in hospital mortality and surgeon. Two distinct models were used. Model 1; two-week visit by teams (10-16) and Model 2 where a team (6-8) stayed 42-50 weeks consecutively. Standardized Mortality Ratios (SMR) were calculated against contemporary multinational data.
Results: Primary operations (trips)/site: Model 1; Sulimaniyah-39 (2), Basra-68 (4), Najaf-72(5), Nasiriyah-115 (7); Model 2; Nasiriyah-577 (78 operating weeks - over 2y 2 m). Total primary operations 871; RACHS-1 distribution and mortality; 1- 102/2 (1.96%), 2- 397/10 (2.53%), 3-241/46 (19.09%), 4-38/16 (42.11%), 5-1/1, 6-1/1, Non-RACHS and ADULT 91/6. Model 1 visits; 36 operating weeks, 294 surgeries, mean cases/week 8.17 ± 1.84. Model 2 visits - 78 operating weeks (over 2y 2 m), 577 surgeries, mean cases/week 7.4 ± 3.73. No significant difference, p = 0.24. Median age of all the patients 4 years 5 months. Age distribution; 0-30 days-39, 31 days-1 year 146 , 1-18 y 605, adults- 75, no Age 6. Median Age RACHS-1 cases (n = 790), 4.0 years. Unadjusted Mortality, Model 1; 36/294 = 12.2%: Model 2; 46/577 = 7.9% (p = .04). The SMR (95% C.I) Model 1;1.75 (1.23-2.42) and Model 2; 1.43 (1.04-1.91). Risk factors for mortality (OR) neonates (7.46), infant (3.64), *(vs 1-18 y). RACHS 3 (9.54), RACHS 4 (36.36) (vs RACHS 1).
Inferences: Model 2 provided more operations and lower SMR secondary to a continuous presence and 3 sustained programs resulted.

P2785 - PROGRAMMATIC CHANGES TO REDUCE MORTALITY AND MORBIDITY IN HUMANITARIAN CONGENITAL HEART SURGERY

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Background: The International Children's Heart Foundation (ICHF) is a non-governmental organization providing congenital cardiac surgery services to the developing world. A review of the organization's outcomes prompted several programmatic changes aimed at improving mortality and morbidity. The purpose of this study is to analyze the impact of those changes on clinical outcomes.
Materials and Methods: In 2015 the organization implemented several quality improvement policies. These included the following: new sites would not accept patients less than 10 kgs, operative complexity would rise only as local capacity increased, preoperative evaluation was to mirror that of the first world (i.e. withholding operative treatment until all medical illness are stabilized, i.e. viral infection), all sites must have a blood bank, medical subspecialty must be available, 24 hour intensivist coverage must be available and a single ICHF surgeon will provide services for each trip. A retrospective review of a prospectively maintained database was conducted, dividing patients into an Early group (2008-2014) and a Late group (2015-2016).
Results: There were 3,521 patients identified in the Early group and 262 in the Late group. Patients in the late group were older and less likely to admitted to the ICU preoperatively, but more likely to be malnourished (Table 1). Bypass and cross clamp times were similar (Table 2). Patients in the Late group had a significantly lower mortality rate (Early 8.5%, Late 2.29%, $p < 0.0001$). The Late group was less likely to undergo reoperation (Early 11.79%, Late 2.29%, $p = 0.0001$) and re-intubation (Early 10%, Late 5.73%, $p = 0.022$) (Table 3). Mortality for the most commonly performed operations also decreased, although not significantly (Table 4).
Conclusions: Programmatic changes undertaken by ICHF has resulted in a significant decrease in mortality and morbidity in the humanitarian setting.

Table 1.

	2008-2014 (n = 3521)			2015-2016 (n = 262)			
Age (Years)	**Value**	**%**	**SD**	**Value**	**%**	**SD**	**P**
Mean	4.96		6.61	6.28		5.9	**0.0007**
Median	2.67			4.64			
Gender (Male)	1816	51.50%		125	47.71%		0.4
Height (cm)	96.72		33.53	105.8		32.6	**0.0001**
Weight (kg)	16.28		14.26	20.1		15.9	**0.0001**
Malnourished	700	19.88%		67	25.57%		**0.03**
Malnourished	558	15.85%		64	24.43%		**0.0005**
Emaciated	142	4.03%		3	1.15%		**0.017**
Preoperative ICU	1003	28.49%		13	4.96%		**0.0001**

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Table 2.

	2008-2014		2015-2016		
	Mean	**StPev**	**Mean**	**StPev**	**P**
CPB (min)	100.28	78.39	101.7	61.8	0.77
Cross Clamp (min)	53.88	38.99	55.2	42	0.6
Intra-Op Blood Products	864 (24.5%)		149 (56.8%)		**0.0001**

Table 3.

	2008-2014		2015-2016		
Re-operation	415	11.79%	6	2.29%	**0.0001**
Bleeding	209	5.94%	5	1.91%	**0.0034**
Revision	98	2.78%	1	0.38%	**0.12**
Pacemaker	20	0.57%	0	0.00%	0.39
Re-intubation	355	10%	15	5.73%	**0.022**
in-hospital mortality	299	8.2%	6	2.29%	**0.0003**

P2809 - PEDIATRIC CARDIAC ASSISTANCE AND DEVELOPMENT IN EASTERN LIBYA 2012 2016

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Background: Prior to the revolution Libya had 1 center performing pediatric cardiac surgery in Tripoli. Following the revolution we were asked to provide pediatric cardiac services and development to Eastern Libya. We began our program in Benghazi in 2012 and were forced secondary to conflict to move to Tobruk in 2015. The results of this program in a region of conflict are presented.
Methods: We queried our database for all trips. Data analyzed were patient demographics, number of trips, primary operations, RACHS Classification, outcomes and surgeon. Models were two-week (Model 1, 14-16 team members) and month-long visits (Model 2, 7-8 team members).
Results: There were 257 males. Median age Benghazi 1.88 yrs, Tobruk 1.27 yrs. Total trips/operations (19/498); weeks/operations, Benghazi (36/260) and Tobruk (27/237). Model 1 mean operations/trip, 20.2 ± 4.4. Model 2 Benghazi, mean operations/trip, 22.71 ± 10.03, Model 2 Tobruk, 34.0 ± 8.3, $p < 0.0001$. Mean operations/week, Model 1, 7.6 ± 1.1; Model 2, 9.8 ± 0.8, $p = 0.009$. RACHS distribution: Benghazi 1-51 (21%), 2- 136 (55.8%), 3- 48 (19.7%), 4- 8 (3.3%), Non-RACHS/ADULT 17, Total 260,. Tobruk 1- 45 (20%), 2- 97 (43.1%), 3- 70 (31.1%), 4- 13 (5.8%), Non-RACHS/ADULT 13, Total 238. Unadjusted mortality: 30/498 (6.02%); RACHS classifiable, Benghazi 10/243 (4.1%), Tobruk 20/225 (8.8%) ($P = 0.03$). RACHS category mortality: Benghazi 1- 0/51 (0%), 2- 2/136 (1.47%), 3- 5/48 (10.42%), 4- 3/8 (37.5%); Tobruk 1- 0/45 (0%), 2- 3/97 (3.09%), 3- 14/70 (20%), 4- 3/13 (23.1%). The SMR Benghazi was 0.804 (0.4-1.4) and Tobruk 1.420 (0.88-2.1). The SMR (95% CI) Model 1 was .64 (.16-1.7) and Model 2 1.24 (.82-1.8). Risk factors for mortality; neonatal age (OR 20.79 = 95% C.I. 6.1-71) and RACHS-1 Category 3 (OR 31.75), and Category 4 (OR 99.9). Local Surgeon primary; Benghazi 91/260, Tobruk 69/238, ($p = .04$).
Inferences: Pediatric cardiac development can be carried out in conflict areas.

P2877 - DEVELOPMENT OF A CARDIAC PEDIATRIC SURGERY PROGRAM AT A PUBLIC HOSPITAL IN LA PAZ BOLIVIA AT 3600 M

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Introduction: Only 20% of bolivians have a health insurance. At high altitude congenital heart diseases are twice as frequent as at sea level. Children with CHD depended til the begin of the program on compasive missions and caritative foundations to get a correction of their diseases. Only 12% were operated in this way and 88% died during the natural evolution.
Aim: To organize a cardiac surgery program in the public children hospital "Hospital del Niño".
Method: a) Infrastrutcture and equipment: We considered key services at the hospital, including children surgery, pediatric ICU, la and blood bank. Modernization of infrastructure and adquisition of equipment were organizaed in consence with the help of several organizationd including the german and japanese cooperation b) Personal qualification: 30 nurses and doctors got a qualification in the postoperative managment of children cardiac surgery. We edited togheter a protocol for every single stage of diagnosis and therapy. Satelite hospitals doctors and nurses were capacitated in the early diagnosis of CHD for opportune reference. c) Political support. Several meetings with authorities of the Ministry of Health conduced to the inclusion of medicaments in the national list of drugs. Steps to include financing of cardiac surgeries are underwent in the office of public insurance.
Conclusion: Allready thirty patients were operated in RACHS categories 1-3, including VSD, ASD, Fallot and palliative surgery for more complex CHD. The mortality up to 30 days is 0%. Children with a CHD and without economical resourses have now an oppotunity to be healed.

P2986 - A STRUCTURED INTERVENTION PROGRAMME CAN IMPROVE THE BIOPHYSICAL WELLBEING IN CHILDREN WITH CONGENITAL HEART DISEASE

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Introduction: Improved survival among children with congenital heart disease (CHD) has shifted focus to long-term physical and psychological outcomes. The benefits of an active lifestyle within the general population have been well described. This study aims to determine if a structured intervention programme can improve

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both physical and psychological functioning in children with CHD.
Methods: This is a prospective randomised control trial. Patients aged 5-10 years with CHD were identified and invited to participate. Each patient underwent baseline assessment as detailed:
Biophysical assessments: • Weight, height, waist measurements • Heart rate, blood pressure, oxygen saturation
• Exercise stress test–Graded cycle ergometer protocol • Actigraph accelerometer worn for 1 week
Psychosocial assessments: • Kidscreen27 • Strengths and Difficulties Questionnaire • Butler Self-image Profile
Following baseline assessment patients were randomised into intervention and control groups. The intervention group attended a one day education session during which motivational interviewing techniques were used to deliver exercise and positive lifestyle advice. They received an individual written exercise plan to take home and implement. The control group continued with their usual level of care. After 4 months all participants were reassessed.
Baseline Results: • 163 patients were recruited, 100 males (61.3%), mean age of 8.4 years (range 5.3–11.5)
• Patient subgroups: 18.4% acyanotic no intervention, 37.4% acyanotic repaired, 27.6% cyanotic corrected, 16.6% cyanotic palliated • EST duration mean 5.89 mins (SD 2.02), • Actigraph: Average time spent in MVPA 45 mins (SD -27.2) • The 'cyanotic palliated' subgroup had significantly shorter EST duration and lower levels of daily MVPA. This subgroup also scored significantly lower on HrQOL physical wellbeing
Review Results: • There was a significant improvement in performance at peak exercise in the intervention group following the exercise programme
Conclusions: Overall physical and psychological wellbeing is well preserved in the majority of children aged 5-11 years with CHD. A structured intervention programme significantly improved peak exercise capacity.

P3002 - CREATING PROFESSIONAL DEVELOPMENT OPPORTUNITIES FOR PEDIATRIC CARDIAC CRITICAL CARE NURSES IN SOUTH VIETNAM

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Nursing education in low and middle income countries (LMICs) still doesn't meet the needs of patients with congenital heart disease. Little formal continuing medical education (CME) is available for pediatric cardiac critical care nurses to advance their skills and knowledge. To address this need, we partnered with the University of Medicine and Pharmacy and the Nursing Teachers Branch in Ho Chi Minh City to develop a CME course for pediatric cardiac critical care nurses. The course was taught by pediatric cardiac care providers from local teaching hospitals and was delivered in two cycles: 2014-2015 and 2015-2016. Each cycle consisted of three, 3-day long sessions taught over a 12 month period. Topics included congenital heart disease anatomy, post-operative management, family-centered care, Basic Life Support and Pediatric Advance Life Support. Teaching approaches included lectures, case studies, scenarios, and group discussions. Pre and post-test scores were collected for each session and the 3 month BLS/PALS follow up. In Year 1, 37 pediatric cardiac critical care nurses from 12 hospitals in the Mekong Delta participated. In Year 2, 21 nurses from 7 hospitals participated. Participant feedback was positive and changes in BLS/PALS preparedness illustrated a significant gain in knowledge and understanding of practices. In Year 1, 19 nurses scored below 60% in the pre-test compared to 10 in the post-test. In Year 2, 14 scored between 60% and 80%, and five scored below 60% in the pre-test. In the post-test, all participants scored at or above 60%. Throughout the course, Children's HeartLink contributed to curriculum development, funding, speaker and participant recruitment. Challenges included participant recruitment (especially outside of Ho Chi Minh City), communication, and long-term follow up to ensure sustained change in practices. An effective subspecialty nursing CME course in LMICs can be developed and implemented when clinical, educational, and

IMAGING

P1034 - HIGH RISK ANGIOGENIC GENES ARE ASSOCIATED WITH ABNORMAL CARDIAC MECHANICS IN CHILDREN WITH HYPERTROPHIC CARDIOMYOPATHY

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Introduction: We have previously reported diastolic dysfunction in children with hypertrophic cardiomyopathy (HCM) associated with high risk (HR) alleles in angiogenic genes (VEGF, HIF1α, and TP53). This study aimed to investigate myocardial deformation and relaxation in high risk (HR) and low risk (LR) genotypes.
Methods: Subjects with HCM (<21yrs) were prospectively enrolled (2007-2010) and genotyped for VEGF (2578 A/C); (1154 A/G), (634 C/G), HIF1α (145 C/T); (1326 C/T), TP53 (97-29 C/A); (Arg72ProC/G) polymorphisms. Presence of an allele associated with down regulation of angiogenic genes was defined as HR genotype. Systolic and diastolic longitudinal, circumferential, and radial strain and strain rate at each myocardial segment were measured by 2D speckle tracking (EchoPac, GE). HR and LR genotypes were compared by Student's t-test.
Results: Fourteen HCM subjects (mean age 6yrs (range 1-15), 79% male, 79% Caucasian) were enrolled. Frequency of HR alleles A, A, T in VEGF were 64%, 50%, 36%, HIF1α (T, C; 71%, 79%) and TP53 (G, C; 64%, 64%) respectively. Septal and posterior wall thickness was similar between LR and HR except for thicker septum in HR VEGF (1154 A/G) ($p = 0.03$) and posterior wall in HR HIF1α (1326 C/T) ($p = 0.08$). In HR genotypes, both systolic circumferential strain and systolic and diastolic circumferential strain rate were significantly decreased in multiple segments, predominantly at the basal and apical levels (i.e. anteroseptal and anterior segments). HR genotypes had significantly decreased systolic longitudinal strain and strain rate at the basal septum, midseptum and basolateral segments, and decreased basal global radial peak systolic strain.
Conclusions: HR alleles that downregulate angiogenic genes are associated with adverse cardiac mechanics in children with HCM.

P1049 - VEIN OF GALEN MALFORMATION WITH DEXTROCARDIA SINGLE LEFT SIDED SVC ABERRANT RIGHT SUBCLAVIAN ARTERY AND INTRACTABLE CARDIAC FAILURE; CHALLENGES IN MANAGEMENT

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Background: Vein of Galen malformation (VOGM) is associated with cardiac anomalies, intractable cardiac failure and signs similar to that of pulmonary hypertension, making management of such cases challenging. We present the challenges we faced in managing a term newborn with VOGM with dextrocardia and single left SVC.
Materials: Madam S was referred to our centre at 33 weeks' gestation for polyhydramnios, abnormal fluid collection in the fetal corpus callosum, and cardiomegaly. Fetal MRI confirmed the presence of VOGM. Fetal echo confirmed dextrocardia, cardiomegaly and dilated left sided SVC. Post delivery echo confirmed the dextrocardia, dilated SVC and cardiomegaly, revealed a large PDA with bidirectional shunting at the PFO and PDA, and tricuspid regurgitation with a pressure gradient of 79 mmHg. She was intubated, ventilated, and started on frusemide. Coil embolisation of 4 feeder vessels to the VOGM resulted in a decrease in SVC diameter and a reduction of ventilator support. Shunting at both PDA and PFO became left to right, and she subsequently developed intractable cardiac failure which remained poorly controlled despite maximising anti failure therapy. Cardiac CT following a failed attempt at ligating the ductus revealed a single left SVC draining into the coronary sinus, a right innominate vein, left sided aortic arch, and an aberrant tight subclavian artery arising from the descending aorta. The child was planned for coil embolisation of a feeder vessel which had increased in diameter. However, baby S became bradycardic and died despite resuscitative efforts during the procedure.
Conclusion: This case highlights the challenges faced in imaging, diagnosing and managing congenital heart disease and cardiac failure in a patient with VOGM. To our knowledge, this is also the first reported case of dextrocardia, single left SVC and aberrant right subclavian artery in a patient with VOGM.

P1084 - HOW DOSE ECHOCARDIOGRAPHY HELPS US BEFORE DURING AND AFTER TRANSCATHETER VENTRICULAR SEPTAL DEFECT CLOSURE PROCEDURE

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Background: Gold standard treatment for ventricular septal defects (VSD) is surgical closure, with minimum mortality but associated risks such as complete AV block (AVB), residual shunt, pericardiotomy syndrome, infection, etc. Transcatheter closure of VSD emerges as an alternative with acceptable morbi-mortality and encouraging results. The purpose of this study is to analyze how echocardiography helps us before, during and after transcatheter VSD closure procedure.
Materials and Methods: A descriptive analysis was performed based on the review of clinical, echocardiographic and catheterization records of patients (p) with transcatheter VSD closure during the period between May 2010 and March 2016 at Pedro de Elizalde Children's Hospital. 51 procedures were performed in 47p. Median age was 7 years old (range 0,33 to 15y). Median weight was 26 Kg (range 4,3-83Kg); 5 procedures were done in p less than 10Kg.
Results: 43 procedures were successful (84,3%), 41 in the first attempt and 2 needed a second procedure. 3p evidenced major complications (6,9%): 1p tricuspid stenosis; 1p hematuria with decrease in blood count ; 1p, <10 kg with muscular infundibular VSD and no aortic prolapse or insufficiency pre procedure, had severe aortic insufficiency immediately post procedure and died. 8 procedures failed in the first attempt, in 2 cases changing the device achieved closure successfully in a second procedure. 6p required surgery (11,7%).
Conclusions: Echocardiography was part of the learning curve and allowed to define fundamental aspects for the correct device selection as VSD anatomic characteristics and its relation with adjacent structures. It's a complementary tool to guide the procedure and provides direct view in key situations as arteriovenous loop, progress, deployment and release of the device, among others. It's essential to assess the correct implantation and stability of the device. It identifies complication during and post procedure and allows decision-making.

P1087 - ISOLATED LEFT COMMON CAROTID ARTERY FROM THE MAIN PULMONARY ARTERY ASSOCIATED WITH TETRALOGY OF FALLOT

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Background: An isolated left common carotid artery (LCCA) is very rare. It would be caused by partial regressions of the aortic sac and the left fourth arch in early fetal life.
Case Report: The fetus was diagnosed as Tetralogy of Fallot (TOF) at 31 weeks' of gestation. She was born at 40 weeks, weighted 3379 g, and 97% in oxygen saturation. The echocardiography and the computed tomography revealed TOF and right aortic arch, from where a right common carotid artery, right subclavian artery, and left subclavian artery branched in order. LCCA was arising from pulmonary artery (PA) via ductus arteriosus (DA). The both sides of brain vessels were communicated with the circle of Willis. With the concern of steal phenomenon due to the connection between LCCA and PA via DA as the pulmonary resistance decreased, the DA was closed at day 1. The SPECT and the cerebral angiography revealed well left brain perfusion. The neurological and developmental status was well at the age of three months. A total correction of TOF was planned in infancy.
Conclusion: LCCA from PA is very rare, and only eight cases have been reported so far. Fong and Venables postulated according to Edwards hypothesis that the distal migration of the left subclavian artery beyond the left ductus arteriosus with subsequent disappearance of two different segments between ascending aorta and LCCA and between left subclavian artery and left DA, which would produce an isolated left common carotid artery and an aberrant left subclavian artery. Our case would support this concept as we could follow the closure of the vessel between LLCA and PA after birth, which was suggestive of ductal tissue.

P1090 - TISSUE DOPPLER VENTRICULAR FUNCTION PARAMETERS CORRELATION WITH CARDIAC BIOMARKERS IN NEWBORNS WITH CONGENITAL HEART DEFECTS

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Background/Hypothesis: Our hypothesis is that tissue Doppler parameters of bi-ventricular function correlate with cardiac biomarkers in newborns with congenital heart defects.
Materials and Methods: Between August 2012 and March 2015, newborns with prenatal diagnosis of congenital heart defects were admitted consecutively to a Neonatal Intensive Care Unit. Healthy newborns delivered in the same hospital were recruited as controls. Plasma levels of cardiac biomarkers (brain natriuretic peptide, creatine kinase-MB, troponin I, myoglobin) were measured in the cord blood, and echocardiograms were performed in both groups.
Results: No significant differences in cardiac biomarkers were observed between the congenital heart defect and control groups. The creatine kinase-MB levels differed by type of congenital heart defect and were significantly higher in cyanotic than in acyanotic congenital heart defects, with median levels of 3.1 (2.6–3.8) and 4.6 (3.2–6.1 ng/mL), respectively, $p = 0.007$; the higher levels were found in the transposition of great arteries group. The creatine kinase-MB cord blood levels positively correlated with the tricuspid valve E/E' ($p = 0.002$; $rho = 0.772$). The troponin I levels negatively correlated with the mitral valve lateral annulus slope ($p = 0.04$; $rho = -0.412$).
Conclusions: The creatine kinase-MB cord blood levels correlated with diastolic ventricular function parameter, while troponin I correlated with systolic function parameter, as evaluated by tissue Doppler.

P1099 - PAPILLARY MUSCLE NONCOMPACTION A UNIQUE MECHANISM FOR MITRAL REGURGITATION

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Introduction: Noncompaction cardiomyopathy (NCCM) is increasingly being reported with variable clinical presentations. We described the first series of patients with noncompaction cardiomyopathy (NCCM) associated mitral regurgitation (MR)in 2004. Subsequently, many other patients were reported by us and others.
Methods: Prospective follow up of cases of NCCM associated with papillary muscle NCCM and MR.
Results: We identified 25 patients with NCCM associated with MR secondary to papillary muscle affection and followed them for 1-10 years (2004-2015). Clinical presentation includes heart failure in 20 patients and a heart murmur without symptoms in 5. Echocardiographic features in all patients showed NCVM involving papillary muscle base with variable degree of chordal elongation /rupture. The mitral leaflets appear retracted and malcoapting. MR degree varied from mild to severe. Ejection fraction was normal >55% in 15 patients, mildly impaired (45-55%) in 5 and severely impaired in 5 patients. Tow patient had mitral valve replacement with good long term outcome.
Conclusion: In NCCM, papillary muscle affection can lead to MR with characteristic echocardiographic features including chordal abnormalities and malcoaptation.

P1113 - DILATION OF THE AORTA OR NEOARTIC ROOT IN THOSE HAVING UNDERGONE FONTAN PALLIATION

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Background: Dilation of the aorta or neoaorta occurs after the Fontan palliation. Cardiac magnetic resonance imaging (CMRI) allows for adequate visualization and measurement of the aorta or neoaorta in this setting. This study was conducted to identify risk factors for such dilation.
Materials and Methods: Patients having had undergone Fontan palliation and a subsequent CMRI study at our institution were identified. CMRI studies were then reviewed with the aorta or neoaorta being measured using a coronal cine image in systole. Measurements were z-scored using values for normal aortas. The median z-score for each anatomic site was then calculated with those having a value above the median being classified as dilated. Univariate analysis was then conducted to identify features associated with dilated aortas.
Results: A total of 84 patients were included. Median z-score was 2.93 (-1.80 to 6.37) for the annulus, 2.13 (-3.56 to 6.66) for the sinus, 2.79 (-2.12 to 8.39) for the ST junction, and 2.53 (-1.59 to 20.00) for the ascending aorta. Aortic insufficiency of at least mild severity was present in 21%, while moderate or severe aortic insufficiency was present in 5%. No risk factors were identified for dilated aortic annulus. Moderate or severe aortic insufficiency was associated with a dilated sinus ($p = 0.03$). History of HLHS, DKS procedure, or a BT shunt were associated with dilation of the ascending aorta.
Conclusions: Factors associated with dilated aortic annulus, sinus, ST junction, and ascending aorta were identified in this study. For the entire cohort, moderate or severe aortic insufficiency was associated with a dilated sinus and history of having undergone a DKS was associated with dilation of the ST junction while having HLHS was associated with dilation of the ascending aorta.

P1114 - IS LV DYSSYNCHRONY AN EARLY MARKER OF VENTRICULAR DYSFUNCTION WITH WORSENING AORTIC REGURGITATION IN YOUNGER PATIENTS WITH CONGENITAL AORTIC VALVE DISEASE CLINICAL APPLICATION OF REAL TIME ECHO DERIVED DEFORMATION INDICES

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Background: Standard echo protocols primarily rely on left ventricular (LV) ejection fraction and shortening fraction to assess ventricular function. Commonly, these parameters are preserved even with severe aortic valve disease. We measured LV deformation during routine clinically-indicated echocardiograms using 2D speckle tracking in patients with congenital aortic stenosis (AS) and/or aortic regurgitation (AR).
Material and Methods: An advanced echo protocol performed at the ultrasound machine and immediately available to caregivers included 2D LV regional and global circumferential (circ) strain (S) and strain rate (SR) from a short axis view and longitudinal (long) S and SR from an apical view, standard deviation of time to peak systolic circ and long S, and max systolic wall delay to peak S was instituted for all patients with AS and/or AR.
Results: 70 consecutive patients (mean age = 16y, range 1 m to 55y; 50 male, 20 female) completed the echo protocol between 12/2014-4/2015. Sixty-two (89%) patients had a bicuspid aortic valve. Ejection fraction was preserved in all patients regardless of aortic valve disease severity (table), and there were no significant differences in circ S, long S, or long SR between patients based on AS or AR severity. In fact, circ S was significantly higher for patients with severe AS. Dyssynchrony was not seen with

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dominant AS disease, but circ standard deviation of time to peak strain and circ max wall delay were significantly higher in patients with severe AR compared to mild-moderate regurgitation.
Conclusion: Both traditional echo assessment and 2D deformation indices suggest preserved LV function in younger patients with congenital aortic valve disease, regardless of severity. Real-time deformation analysis does identify progressive dyssynchrony with worsening AR, however, which may be marker of early LV dysfunction. This technique should be added to routine clinical surveillance.

P1115 - SEGREGATING BODILY ISOMERISM POTENTIAL ECHOCARDIOGRAPHIC CORRELATIONS OF MORPHOLOGIC FINDINGS

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Background: Bodily isomerism, also referred to as heterotaxy, involves predominantly the thoracic organs, although other organs are usually abnormally positioned. Previously assessed on the basis of splenic anatomy, it is now understood that isomerism is better segregated on the basis of atrial appendage morphology. This allows for anticipation of associated findings. We aimed to assess the accuracy of segregation based on the morphology of the atrial appendages and other structures more easily identified by echocardiography.
Materials and Methods: We reviewed postmortem specimens of hearts from the archives at four institutions categorized as obtained from patients with "heterotaxy". The cardiac structures were analyzed using sequential segmental analysis. Non-cardiac structures were also examined if available. Statistical analysis was performed to compare differences in the settings of right as opposed to left isomerism.
Results: Specimens were available from 188 patients. Of these, 57 had left isomerism, and 131 had right isomerism. Atrial appendages were isomeric in all patients. A coronary sinus was found only in left isomerism, while a terminal crest, or a Eustachian valve, was only in right isomerism. Interruption of the inferior caval vein was associated with left isomerism, while totally anomalous pulmonary venous connection was associated with right isomerism.
Conclusion: Isomerism is uniformly segregated on the basis of the morphology of the atrial appendages, itself defined by the extent of the pectinate muscles. These features themselves, if knowingly sought, should be identifiable in the clinical setting, but other cardiac findings can be used for segregation by the echocardiographer.

P1117 - VESTIBULAR ATRIAL SEPTAL DEFECTS A NECROSCOPY STUDY

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Background: The vestibular atrial septal defect is a newly described type of interatrial communication that is a defect in the antero-inferior portion of the atrial septum. A result of inadequate muscuiarization of the vestibular spine and mesenchymal cap, the vestibular atrial septal defect represents a true deficiency of the atrial septum. A review of necroscopy specimens from three separate archives was conducted to further characterize vestibular atrial septal defects and associated cardiac findings.
Materials and Methods: Sequential segmental analysis was utilized to analyze hearts in the Farouk S. Idriss Cardiac Registry at Ann & Robert H. Lurie Children's Hospital in Chicago, IL, the Van Mierop Archive at University of Florida in Gainesville, Florida, and the archive at Johns Hopkins All Children's Heart Institute in St. Petersburg, Florida. Hearts with a vestibular atrial septal defect were identified and further characterized to find findings associated with such atrial septal defects.
Results: A total of 2,100 specimens were analyzed. Of these, 68 (3%) were found to have a vestibular atrial septal defect. Such defects were associated with bilateral atrial dilation, fossa ovalis type atrial septal defects, abnormalities of the right sided atrioventricular valve, pulmonary atresia, right ventricular outflow tract obstruction, and the presence of an interventricular communication. Concordant atrioventricular and ventriculo-arterial junctions were present in nearly all cases.
Conclusion: The vestibular defect, therefore, is a recently recognised true atrial septal defect located in the muscular antero-inferior rim of the oval fossa. Our retrospective review of autopsied hearts suggests that the defect may be commoner than previously thought. Diagnosis represents an echocardiographic challenge, as the location of the defect can mimic a multi-fenestrated floor of the oval fossa. Increased awareness of the defect should optimize its future clinical identification.

P1131 - DOUBLE AORTIC VALVE A PREVIOUSLY UNREPORTED ANOMALY

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Background: We present a previously unreported congenital heart anomaly with duplications of aortic valve, aortic sinus and proximal ascending aorta.
Materials & Methods: A six years old girl presented with complains of cyanosis and effort intolerance. On trans-thoracic echocardiography (TTE), she was diagnosed with Double-outlet right ventricle (DORV), ventricular septal defect (VSD) and infundibular stenosis. Surgical correction was scheduled with a plan of routing the VSD to the aorta and resection of infundibular stenosis.
Results: During surgery, the aortic root appeared significantly enlarged which tapered to a normal looking ascending aorta giving a "flask like" appearance. The right atrium was opened after instituting the cardiopulmonary bypass and cardioplegic arrest. In addition to a large sub aortic VSD and infundibular stenosis, the aortic root appeared to be guarded by two seperate aortic valves, oriented antero-posteriorly and separated by a muscle bar. The anterior aortic valve was tricuspid and posterior aortic valve was bicuspid. The VSD was routed to the aorta committing both aortic valves to the left ventricle and infundibular obstruction was resected. The postoperative course was uneventful and hospital discharge occurred on fifth postoperative day. Computed tomography revealed duplications of aortic valve, aortic sinus and

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proximal ascending aorta. The coronaries arose from the posterior sinus. *Conclusions:* The distal part of the embryological bulbous cordis forms the roots and proximal portions of aortic and pulmonary artery. Duplication occurring at this stage could lead to such an anomaly, although it has never been reported before. We present the first-ever case of duplicated aortic valve, aortic sinus and proximal ascending aorta.

P1132 - LEFT VENTRICULAR OUTFLOW TRACT ANEURYSM FOLLOWING TRAUMA

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Background: Congenital or acquired cardiac aneurysms are exceedingly rare in children. Acquired aneurysms may occur due to trauma, ischaemia, infection or iatrogenic injury. We present the rare case of traumatic aneurysm involving left ventricular outflow tract (LVOT).
Materials & Methods: A 6 year-old girl sustained fractures of her left humerus and left hip, and blunt trauma to her left chest after a fall. Her fractures were treated surgically. Six weeks later, transthoracic echocardiography (TTE) performed for diagnostic evaluation of fevers, revealed a pericardial effusion and a cystic mass in the transverse sinus. Subsequent Computerised tomography (CT) showed an aneurysm arising from the LVOT which occupied most of the transverse sinus, compressing the proximal right pulmonary artery, left sided pulmonary veins and the left atrium. The mouth of the aneurysm measured 10*10mm and was located 2mm below the aortic annulus.
Results: Surgical repair was performed through median sternotomy on cardiopulmonary bypass. The Aorta was opened transversely after aortic cross-clamp and cardioplegic arrest. Aortic valve leaflets were retracted to expose the LVOT and locate the aneurysm mouth. The defect was closed with a Darcon patch without disturbing the actual aneurysm. The postoperative TTE revealed a significantly shrunk aneurysm with spontaneous echo contrast. There was no residual communication between the left ventricle (LV) and the aneurysm. The postoperative course was uneventful, requiring mechanical ventilation for 3 hours and hospital stay of 5 days.
Conclusions: Traumatic aneurysms of LV following blunt chest trauma are exceedingly rare at any age and especially so in children. The probable cause is injury to coronary artery branch or contusion of myocardium leading to scarring, thinning of scar tissue and aneurysm formation.

P1135 - ANOMALOUS LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY THE ROLE OF ECHOCARDIOGRAPHY

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Objective: Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare but potentially life-threatening congenital heart defect, whose detection still remains a challenge. A retrospective analysis was made to elucidate the role of echocardiography in ALCAPA.
Methods: Fifteen consecutive patients, aged from 1 month to 7 years, received the surgery of ALCAPA between August 2012 and October 2016, were included. Echocardiography was carried out in all before surgery, and selective coronary angiography was performed in 7 cases.
Results: (1) In our series, 14 cases were diagnosed as ALCAPA accurately by echocardiography before surgery. Among which, 3 cases was misdiagnosed as dilated cardiomyopathy and 1 case as severe mitral regurgitation by initial echocardiography, all of the 4 cases were corrected as ALCAPA by echocardiography during the follow-up. Another one case was misdiagnosed as coronary artery dilation, which was corrected as ALCAPA by selective coronary angiography. (2) The infant type was found in 13 cases with few collateral vessels manifested as cardiomegaly, left ventricular dysfunction and mitral regurgitation, and the adult type was in 2 cases with abundant collateral vessels remained normal heart size and better cardiac function. (3) After left coronary artery transplantation, 2 cases died within 1 month due to heart failure, and another 13 cases were followed-up for (22.8 ± 15.3) months. The left ventricular ejection fraction increased from (49.4 ± 12.1) % to (64.9 ± 9.1) % ($t = 3.467$, $p = 0.007$). Left ventricular end diastolic diameter decreased to normal range in 9 cases, and the other 4 cases were still enlarged slightly. Functional mitral regurgitation also reduced in degree.
Conclusion: Echocardiography plays an important role during the perioperative period of ALCAPA. The combination of cardiomegaly, left ventricular dysfunction and mitral regurgitation is the diagnostic clue, however, a normal heart size and cardiac function cannot rule out the diagnosis.

P1138 - QUANTITATIVE ASSESSMENT OF LEFT VENTRICULAR ROTATION AND TWIST USING MAGNETIC RESONANCE IMAGING IN PATIENTS WITH REPAIRED TETRALOGY OF FALLOT

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Purpose: Early detection of left ventricle (LV) dysfunction is very important for evaluating clinical outcome in patients with repaired tetralogy of Fallot (TOF). The index of LV ejection fraction (LVEF) may underestimate the degree of deteriorated cardiac function. In patients with repaired TOF, abnormal LV twist by echocardiography has been demonstrated, but there is still no report about LV rotation and twist by magnetic resonance imaging (MRI). We aimed to evaluate LV rotation and twist using MRI in patients with repaired TOF.
Methods: This study consisted of 42 patients with repaired TOF (mean age 22.1 ± 4.8 yrs) and 38 normal subjects (mean age 22.1 ± 1.8 yrs). They all underwent MRI. The volumetric indices, mass, and ejection fraction of both RV and LV were determined. The VØ peak-to-peak (PTP) and the rotation angle, characterizing the twist function of each slice in the LV, were computed. MRI derived parameters were compared between two groups.
Results: Compared to normal controls, patients with repaired TOF had significantly higher RVESVI (62.6 ± 23.0 vs 40.4 ± 9.7 cm3/m2, $p < 0.01$), RVEDVI (120.7 ± 33.2 vs 77.7 ± 11.2 cm3/m2, $p < 0.01$), and RVSVI (56.3 ± 14.3 vs 37.3 ± 7.1 cm3/m2, $p < 0.01$). There was no significant difference between two groups in terms of LVEF (73.2 ± 5.6 vs 70.9 ± 4.5, $p > 0.05$) and RVEF (49.1 ± 6.9 vs 48.1 ± 7.5, $p > 0.05$). Patients with repaired TOF

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showed a significantly lower VØ PTP (2.7 ± 1.3 vs 5.4 ± 1.6 cm/s, $p < 0.01$) and LV rotation angle (4.9° ± 2.8° vs 8.7° ± 2.4°, $p < 0.01$) than normal controls.
Conclusions: Our results demonstrate that lower LV rotation and twist can provide early information about regional abnormalities of LV before a global ventricular dysfunction in patients with repaired TOF. Assessment of LV rotation and twist using.

P1154 - SEMIAUTOMATIC THREE DIMENSIONAL CT VENTRICULAR VOLUMETRY IN PATIENTS WITH CONGENITAL HEART DISEASE AGREEMENT BETWEEN TWO METHODS WITH DIFFERENT USER INTERACTION

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Background: To assess agreement between two semi-automatic, three-dimensional (3D) computed tomography (CT) ventricular volumetry methods with different user interactions in patients with congenital heart disease.
Materials and Methods: In 30 patients with congenital heart disease (median age 8 years, range 5 days–33 years; 20 men), dual-source, multi-section, electrocardiography-synchronized cardiac CT was obtained at the end-systolic (n = 22) and/or end-diastolic (n = 28) phase. Nineteen left ventricle end-systolic (LV ESV), 28 left ventricle end-diastolic (LV EDV), 22 right ventricle end-systolic (RV ESV), and 28 right ventricle end-diastolic volumes (RV EDV) were successfully calculated using two semi-automatic, 3D segmentation methods with different user interactions (high in method 1, low in method 2). The calculated ventricular volumes of the two methods were compared and correlated. A P value <0.05 was considered statistically significant.
Results: LV ESV (35.95 ± 23.49 ml), LV EDV (88.76 ± 61.83 ml), and RV ESV (46.87 ± 47.39 ml) measured by method 2 were slightly but significantly smaller than those measured by method 1 (41.25 ± 26.94 ml, 92.20 ± 62.69 ml, 53.61 ± 50.08 ml for LV ESV, LV EDV, and RV ESV, respectively; $P < 0.02$). In contrast, no statistically significant difference in RV EDV (122.57 ± 88.57 ml in method 1, 123.83 ± 89.89 ml in method 2; $P = 0.36$) was found between the two methods. All ventricular volumes showed very high correlation ($R = 0.978$, 0.993, 0.985, 0.997 for LV ESV, LV EDV, RV ESV, and RV EDV, respectively; $P < 0.001$) between the two methods.
Conclusion: In patients with congenital heart disease, 3D CT ventricular volumetry shows good agreement and high correlation between the two methods, but method 2 tends to slightly underestimate LV ESV, LV EDV, and RV ESV.

P1155 - ATRESIA OF THE BILATERAL PULMONARY VEINS A RARE AND DISMAL ANOMALY IDENTIFIED ON CARDIAC CT

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Background: Imaging findings of bilateral pulmonary vein atresia have not been described. To describe cardiac CT findings and clinical outcomes of bilateral pulmonary vein atresia.
Materials and Methods: Three newborns with bilateral pulmonary vein atresia were encountered at our institution during a period of 8 years. We evaluated prenatal echocardiographic findings, clinical presentations, postnatal echocardiographic findings, chest radiographic findings, cardiac CT findings and clinical outcomes.
Results: All newborns presented immediately after birth with severe cyanosis, respiratory distress and acidosis that were unresponsive to medical management. Prenatal and postnatal echocardiographic studies and chest radiography were misleading, inconclusive or nonspecific in making the diagnosis in these children; however cardiac CT clearly demonstrated atresia of the bilateral pulmonary veins with multiple small mediastinal collateral veins and pulmonary edema. Surgical treatments were not feasible for this anomaly. Their clinical outcomes were universally dismal and all infants died within 3 days.
Conclusion: Cardiac CT provides an accurate diagnosis of bilateral pulmonary vein atresia and leads to prompt treatment decision in these children.

P1201-3D PRINTED MODELS FOR SURGICAL PLANNING IN COMPLEX CONGENITAL HEART DEFECTS AN INTERNATIONAL MULTICENTER STUDY

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Introduction: Three-dimensional printed models (3D-models) provide an unrivalled spatial appreciation of patient-specific cardiovascular structures. However, the impact of this technology on surgical planning in complex congenital heart disease (CHD) is yet to be demonstrated.
Methods: A prospective, observational, case-crossover study involving 10 international centres and 40 patients with complex CHD (median age 3 years, range 1 month- 34 years) was conducted. Magnetic resonance imaging and computed tomography were used to acquire and segment the 3D cardiovascular anatomy. Models were fabricated by fused deposition modelling of polyurethane filament. We sought to evaluate (1) 3D-model dimension accuracy by comparison with medical images, (2) utility of the 3D models by evaluation of subjective satisfaction questionnaire and (3) incremental diagnostic value of 3D-models to plan surgery, comparing decisions made in two scenarios: During routine clinical practice and after inspection of a 3D-model.

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Results: 3D models accurately replicate anatomy with a mean bias of -0.27 ± 0.73 mm. 13 surgeons and 30 paediatric cardiologists completed a satisfaction survey. 96% agree or strongly agree that 3D models provided better understanding of CHD morphology and reduced the chance of complications. 3D models changed the surgical decision in 45% of the cases. Consideration of a 3D-model refined the planned biventricular repair, achieving an improved surgical correction in 20% of cases. In 15% of cases initially considered for conservative management or univentricular palliation, inspection of the 3D-model enabled successful biventricular repair.
Conclusions: 3D models are accurate replicas of the cardiovascular anatomy and improve the understanding of complex CHD. This results in a change in the surgical decision making in 45% of the cases, allowing a better refinement of surgical strategy and surgical correction in cases that would conventionally be considered technically infeasible.

P1214 - IMPROVED ANTENATAL DETECTION AND 30 DAY MORTALITY IN CRITICAL CONGENITAL HEART DISEASE WITH A TWO VENTRICLE OUTCOME A POPULATION BASED STUDY

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Introduction: Isolated reports from selected populations suggest early diagnosis of critical congenital heart disease (cCHD) may result in improved outcome, especially in conditions where the surgical outcome is excellent. However, population-based data are sparse, and their interpretation frequently complicated by the inclusion of complex heart disease and noncardiac comorbidity that may independently influence outcome.
Methods: We undertook a population-based review of cCHD (CHD resulting in operation or death by 30 days) born in New Zealand between 2006 and 2014. Cases were acquired from the National Fetal Cardiology and Cardiac Surgical databases and from the statutory reporting body for perinatal deaths. All diagnoses from 20 weeks gestation and diagnoses made at post-mortem were included. The timing of diagnosis, survival to cardiac surgery and 30-day mortality were reviewed in all livebirths where there was potential for a two-ventricle circulation. Those with a syndrome or additional major noncardiac anomaly were excluded from the analysis as were those born prematurely (<35 weeks gestation).
Results: Of the 436 infants born with cCHD and a potential 2 ventricle circulation, 371 did not have a syndrome or a major non-cardiac abnormality. The rate of antenatal diagnosis increased during the study period ($p = 0.006$) while 30 day mortality declined from 7.0% to 0.9% ($p = 0.049$). Mortality was largely confined to those who died prior to cardiac surgery (Figure). Termination of pregnancy was uncommon and did not vary over the study period.
Conclusions: There has been a significant increase in the rate of antenatal diagnosis, concomitant with a decrease in 30 day mortality in infants with readily treatable cCHD. Earlier recognition of cCHD resulting in appropriate delivery location and a reduced risk of circulatory collapse has very likely contributed to the improved survival.

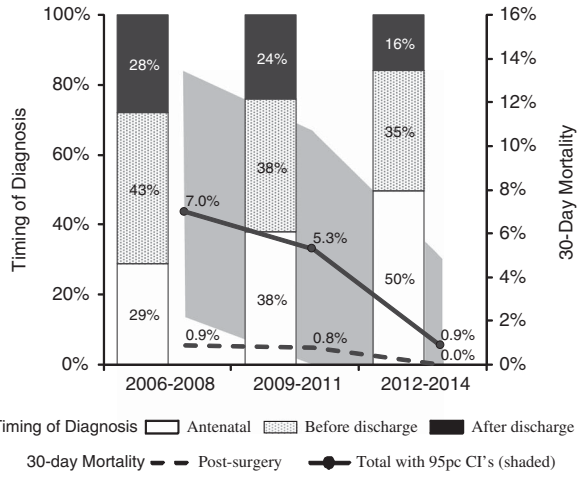

Figure.

P1216 - A RARE CASE OF RIGHT VENTRICULAR NON COMPACTION OR AN EARLY PRESENTATION OF ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA

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Ventricular non-compaction is a rare type of cardiomyopathy that results from arrest of myocardial development and cases in which the right ventricle is involved are extremely rare. We present the case of a 13-year-old boy observed for the presence of premature ventricular complexes (PVC) detected during the Master step test used for the clearance for competitive sport activity. The boy was a young football player. No symptoms were referred and family history was negative for sudden cardiac death or relevant arrhythmias. The ECG at rest was normal and didn't show major abnormalities in ventricular repolarization. During a maximal exercise test on the treadmill (Bruce protocol) we detected rare presence of PVC (with a morphology of left bundle branch block) only in the recovery period. We performed a 24 hours Holter monitoring that showed sporadic PVC. The echocardiogram showed a non-compaction of the right ventricular which it appeared also slightly hypertrophic predominantly in the apical areas. There was no evidence of other cardiac abnormalities associated. In our case the criteria for diagnosis of ventricular non compaction (end-systolic ratio of the non-compacted to compacted myocardial layer >2) were not reached. The presence of the mild structural alterations of the right ventricle and arrhythmias never described before, could suggest an initial form of arrhythmogenic right ventricular dysplasia (ARVD). The age of the patient is compatible with the appearance of structural abnormalities of the myocardium in the ARVD. A cardiac MRI is scheduled for a better structural definition of the myocardium abnormalities and a close follow-up is mandatory. In any case, the screening for physical activity has been shown extremely useful for an early detection of a possible life-threatening cardiomyopathy.

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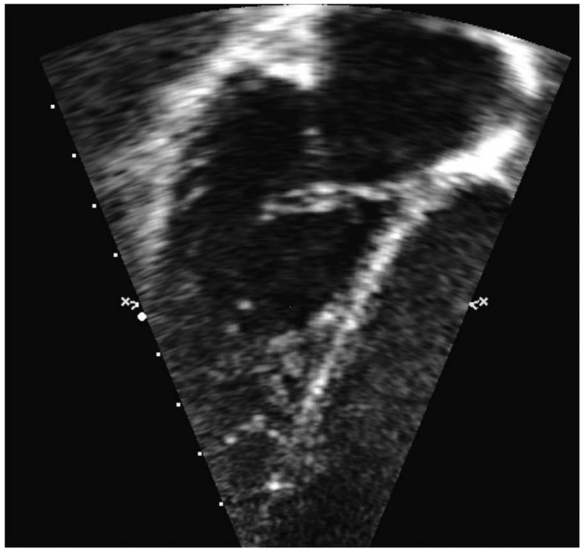

Figure.

P1219 - THREE DIMENSIONAL ECHOCARDIOGRAPHY RIGHT VENTRICULAR INDICES ASSESS DISEASE SEVERITY IN PEDIATRIC PULMONARY HYPERTENSION

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Background: World Health Organization (WHO) functional class is used to assess pulmonary hypertension (PH) disease severity and identified as a treatment goal that correlates with long term outcomes in PH. Maintaining right ventricular (RV) function is important in the survival of PH patients. Three-dimensional echocardiography (3DE), without geometric assumptions, is more accurate than two-dimensional echocardiography in evaluating RV size and function. We used 3DE RV indices to evaluate pediatric PH patients in different clinical status and assessed disease severity.
Methods: 172 children with PH (mean age 9.4 ±5.6yrs; 51% male; WHO: 1-4) underwent 3DE evaluation of the RV from 2014-2016. Offline analysis on the TomTec 4D RV-Function 2 generated RV indices: 3D end-diastolic volume (EDV), 3D end-systolic volume (ESV), 3D ejection fraction (EF), RV septal and free wall longitudinal strain (LS), tricuspid annular plane systolic excursion (TAPSE), and fractional area change (FAC). Kruskal-Wallis and ANOVA were used to evaluate WHO intergroup variabilities in RV indices. Receiver Operating Characteristics (ROC) analyses were performed to identify the best cut-offs in predicting disease severity between WHO I + II and WHO III + IV.
Results: Patients were classified based on their WHO functional class (I = 78, II = 64, III = 20, IV = 10). Significant differences were found in RV EDV, ESV, EF, and free wall LS between WHO 1-4 classes (all $p < 0.0001$). 3D RV EF is the most sensitive parameter to detect worse WHO functional class. Table 1 demonstrates the ROC analyses.
Conclusions: 3D RV volumes, EF, free wall strain, and FAC correlate with disease severity as assessed by WHO functional class. Future studies should evaluate RV functional indices as echocardiographic targets to achieve treatment goals in pediatric PH patients.

Table 1. Right Heart Indices from ROC Analyses

	AUC	Sensitivity	Specificity	Cut-off
EDV (ml)	0.76	61.3	82.9	137
ESV (ml)	0.78	77.4	66.5	52
EF (%)	0.79	90	65.5	43.7
Septal LS (%)	0.65	35.4	93.5	-8.0
Free wall LS (%)	0.72	54.8	91.7	-12.4
TAPSE (mm)	0.52	24.3	91.3	17.9
FAC (%)	0.78	87.1	68.8	34.1

P1239 - THREE DIMENSIONAL ECHOCARDIOGRAPHIC GUIDANCE OF RIGHT HEART CATHETERIZATION DECREASES RADIATION EXPOSURE IN ATRIAL SEPTAL DEFECT CLOSURES

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Background: Radiation dose reduction is desirable in children undergoing cardiac catheterizations. Three-dimensional echocardiography (3DE) provides wide field of view and depth of cardiac structures that cannot be seen in two-dimensional echocardiography. 3DE is used in atrial septal defect (ASD) closure and other interventions. The utility of 3DE in guiding right heart catheterization (RHC) without fluoroscopy for ASD closures has not been demonstrated. We used 3DE guidance of RHC for closure of ASDs and evaluated the radiation reduction compared to historical controls.
Methods: 3DE guidance of RHC and ASD closure was performed in children who were of adequate size to fit the 3D transesophageal probe. Live 3D, in the mid-esophageal view at 90°, was used to guide the catheter from inferior vena cava to the superior vena cava. Using 3D Zoom, the ASD was seen from the right atrial (RA) view and the catheter crossed into the left atrium. Using live 3D at the mid-esophageal view at the 0° and rotation of the 3D probe to a left pulmonary vein, the catheter was guided into the left pulmonary vein. Lastly, using live 3D at the mid-esophageal view at 60° with increased maximal lateral width, the RA, right ventricle (RV), and pulmonary arteries were visualized. The catheter was guided from the RA to the RV, then into the main pulmonary artery. ASD closure was performed with 3DE guidance. Fluoroscopy was used sparingly for device advancement and delivery. Fluoroscopy time and radiation was compared to controls in 2015.

Table 1. Patient demographics, fluoroscopy time, and radiation dose.

Patients	Age (yrs)	Weight (kg)	Height (cm)	BSA	Fluoroscopy (min)	Radiation (mGy)	Device
1	5	21.6	117.3	0.84	3.2	3.22	20 mm GSO
2	7	17.8	115	0.75	3.6	2.5	25 mm GSO
3	11	35.2	136.2	1.15	13.2	8.9	20 mm GSO
4	17	55.8	165.3	1.61	3.5	4	25 mm GSO
5	16	70.9	164	1.77	5	9	25 mm GSO
6	16	67.8	169.2	1.78	1.7	3.5	24 mm GSO
7	6	22	116.4	0.84	4.3	2	25 mm GSO

GSO = Gore Septal Occluder

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Results: 7 patients underwent 3DE guidance of RHC and ASD closures in 2016 (Table 1). There was 67% reduction in fluoroscopy time and 87% radiation reduction demonstrated in 3DE guidance of ASD patients compared to controls (Table 2).
Conclusions: 3DE guidance of RHC and ASD closures resulted in significant reduction in radiation use in children.

Table 2. Fluoroscopy and radiation reduction between controls and 3DE guidance.

	Mean Fluoroscopy (Min)	Mean Radiation (mGy)
2DE ASD controls in 2015	14.5	35.4
3DE Guidance ASD Closures in 2016	4.9	4.7
% Reduction	67%	87%

P1243 - IMPAIRED ELASTIC PROPERTY OF ASCENDING AORTA IN FUNCTIONAL SINGLE VENTRICLE BEFORE FONTAN OPERATION

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Background: Aortic dilatation and impaired elastic properties has been reported as aortopathy in patients after Fontan operation. However, it is little known whether these abnormalities are primary or secondary present. Objectives: We hypothesized that aortic wall abnormalities were primary present in functional single ventricle (SV) before Fontan operation, as well as bicuspid aorta and coarctation of aorta.
Methods: We measured the diameter of aorta at 2 different levels (Ascending aorta [As -AO] and descending aorta [d-AO] by 2D echocardiography. Aortic stiffness index, distensibilty were calculated using these diameters and simultaneously measured right arm blood pressure. Seven neonates with functional SV with pulmonary stenosis or atresia (SV group: media age, 1 day) were compared with 7 normal neonates (Control group: median age, 0 day).
Results: Ascending aortic diameter was greater in patients than that in control (Z sore, SV: 1.3 ± 1.1 vs. Control: -0.1 ± 0.6, $p = 0.04$), but descending aortic diameter was similar between groups. Distensibility of As-AO was reduced (SV: 6.4 ± 2.3 mmHg-110-3 vs. Control: 12.1 ± 4.3 mmHg-110-3, $p = 0.009$) and stiffness index of As-Ao was higher in SV group compared to control group (SV: 5.9 ± 1.1 vs. Control 3.0 ± 1.0, $p = 0.01$). Elastic properties of the d-AO were similar between the groups.
Conclusions: Elastic properties are impaired in neonates who have functional SV with dilated ascending aorta. These abnormalities may be primary present before Fontan operation.

P1260 - BIVENTRICULAR FUNCTIONAL PATTERN IN FETUSES WITH CONGENITAL AORTIC COARCTATION

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Background: Neonates and adults with congenital aortic coarctation (CAC) have been shown to present persistent ventricular dysfunction. In fetal life, CAC induces significant right ventricular dominance, however associated biventricular functional pattern has not been fully described.
Methods: A comprehensive echocardiography was performed at third trimester of pregnancy in 21 fetuses with suspected CAC (2011-2016) in which diagnosis was postnatally confirmed. Two gestational-age-matched controls were selected for each case. Gestational age (GA), estimated fetal weight (EFW) and standard morphometric and Doppler parameters were obtained. Biventricular systolic and diastolic function was assessed by M-mode, pulse-wave and tissue Doppler imaging (TDI) techniques, including: stroke volume (SV), cardiac output (CO), peak ventricular filling velocities and ratio (E/A ratio), mitral and tricuspid annular displacement (MAPSE and TAPSE) and peak annular velocities (S',E',A' and E/E' ratio).
Results: GA and EFW at echocardiography were similar between groups (31.2 vs. 32.2, $p = 0.341$). Fetuses with CAC showed a higher tricuspid to mitral ratio (1.5 vs. 1.03, $p = 0.001$) with significant tricuspid regurgitation and reverse flow at the aortic isthmus in 27% and 25% of cases, respectively. While left function was mainly preserved, the right ventricle showed both diastolic and systolic dysfunction with significantly decreased E/A (0.69 vs. 0.77, $p = 0.028$)) and E/E' ratios (5.1 vs. 5.9, $p = 0.042$), increased E' (9.21 vs. 7.69, $p = 0.006$) and S' (8.02 vs. 7.37, $p = 0.037$) and increased stroke volume (5.37 vs. 3.27, $p = 0.003$) and cardiac output (421 vs. 226, $p < 0.001$) compared to controls. Surgery was performed, on average, at the 7th day of life, with no major complications. Results are summarized in table 1.
Conclusions: In the third trimester of pregnancy, right ventricular dominance in fetuses with CAC is associated with increased right cardiac output, higher longitudinal systolic function and abnormal diastolic function in the right ventricle. Despite a lower stroke volume, left ventricular functional parameters remain relatively preserved.

Table 1.

	CAC Mean (SD)	Controls Mean (SD)	P
Perinatal outcome			
Gestational age at birth (weeks)	40.05 (2.61)	38.12 (3.46)	0.020
Birthweight (g)	3230 (455)	3248 (654)	0.900
Standard fetal Doppler			
Cerebro-placental ratio (MCA PI/UA PI)	1.75 (0.32)	2.19 (0.64)	0.001
DVPI	0.56 (0.16)	0.54 (0.16)	0.609
Right ventricle funcional parameters			
E/A	0.69 (0.12)	0.77 (0.08)	0.028
TDI E' (cm/s)	9.21 (2.74)	7.69 (1.48)	0.006
TDI A' (cm/s)	12.08 (2.04)	11.07 (1.95)	0.075
E/E'	5.12 (1.05)	5.9 (1.33)	0.042
TAPSE (mm)	7.47 (1.76)	6.85 (1.87)	0.215
TDI S' (cm/s)	8.02 (1.13)	7.37 (1.12)	0.037
Stroke Volume (ml)	5.37 (2.62)	3.27 (1.4)	0.003
CO by EFW (ml/min/kg)	421 (93)	226 (70)	< 0.001
Ejection fraction (%)	63.15 (10.89)	57.8 (9.68)	0.105
Left ventricle functional parameters			
E/A	0.76 (0.14)	0.78 (0.11)	0.493
TDI E' (cm/s)	6.85 (1.18)	6.99 (1.32)	0.701
TDI A' (cm/s)	8.77 (1.4)	9.11 (2.06)	0.496
E/E'	6.56 (1.97)	5.64 (1.59)	0.061
MAPSE (mm)	4.93 (1.17)	5.01 (1.27)	0.801
TDI S' (cm/s)	6.81 (1.22)	6.84 (1.35)	0.941
Stroke Volume (ml)	1.84 (1.01)	2.78 (1.56)	0.007
CO by EFW (ml/min/kg)	164 (108)	188 (75)	0.321
Ejection fraction (%)	73.12 (11.07)	67.84 (10.43)	0.097

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P1264 - IMPROVEMENT OF HYPERECHOGENIC MYOCARDIUM AFTER PRENATAL TREATMENT WITH DEXAMETHASONE IN FETUS EXPOSED TO MATERNAL ANTI SSA RO AND ANTI SSB LA ANTIBODIES

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Introduction: Immune-mediated cardiac disease (IMCD) occurs when maternal anti-SSA/Ro and anti-SSB/La antibodies transplacentally damage a developing fetal heart. IMCD is characterized by congenital heart block (CHB), valve disease, cardiomyopathy, and endocardial fibroelastosis (EFE). The mortality of immune-mediated CHB is approximately 20%. Appropriate treatment to prevent IMCD has not been standardized.
Case Presentation: A 26-year-old G1P0 at 24 weeks estimated gestational age (EGA), with no symptoms consistent with systemic lupus erythematosus or Sjogren's Syndrome, had a screening fetal ultrasound notable for markedly hyperechogenic myocardium. Subsequent labs demonstrated elevated titers of anti-SSA/Ro (>8), anti-SSB/La (5.4), and ANA (1:2560). Fetal heart rate was normal (152) with a normal mechanical A-V (PR) interval (106-118 ms). Hyperechogenic foci were noted throughout the heart (bilateral ventricular myocardium, papillary muscles, atrioventricular valves, and atria) raising concern for developing EFE. Cardiac anatomy and systolic function were normal.
Management and Outcome: Given the concern for developing progressive cardiac disease with the presumption that the echogenicity represented injury mediated by autoantibodies, maternal treatment with oral dexamethasone was initiated at 4 mg daily. At 33 weeks EGA, a follow-up fetal echocardiogram revealed decreased myocardial involvement with continued normal ventricular function and A-V intervals. Dexamethasone was weaned to 1 mg daily until successful, term delivery of a baby boy (3.4 kg) with no evidence of rash or distress. Newborn electrocardiogram revealed normal sinus rhythm (rate 101) with normal PR interval (118 ms). Follow-up echocardiograms (last at 11 months of age) showed mild persistent hyperechogenicity of the atrioventricular valves, papillary muscles, and chordae but otherwise, normal valve and ventricular function. Following delivery, both mother and baby underwent successful steroid wean.
Conclusion: Studies of medications to prevent or treat IMCD have included corticosteroids, intravenous immunoglobulin, and hydroxychloroquine. This report describes a case using dexamethasone with favorable outcomes.

P1285 - SPECKLE TRACKING ECHOCARDIOGRAPHY TO PREDICT COARCTATION OF THE AORTA IN NEONATES WITH PATENT DUCTUS ARTERIOSUS

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Background: Coarctation of the aorta (CoA) is challenging to diagnose either in fetuses or in neonates with patent ductus arteriosus (PDA). Speckle tracking echocardiography (STE) allows reliable analysis of myocardial deformation in newborns and seems to provide important insides into regional changes in patients with left ventricular (LV) outflow tract obstruction. The aim of the study was to assess the interest of STE for predicting CoA in neonates with PDA.
Material and Methods: Prospective single-center study in a pediatric cardiac intensive care unit. 28 newborns with either prenatal (n = 12) or postnatal (n = 16) suspicion of CoA and PDA were included. All newborns were evaluated by conventional echocardiography and LV STE. To assess the reproducibility of STE, 14 healthy full-term newborns with PDA were screened. CoA was diagnosed when PDA closed, according to usual echocardiographic criteria.
Results: 11 neonates developed a CoA after PDA closure. Mean gestational age and birth weight were 39.1 ± 1.8 weeks and 3.3 ± 0.62 kg (versus 39.1 ± 1.1 weeks and 2.9 ± 0.60 kg for newborns without CoA, $p = 0.407$ and $p = 0.063$ respectively). STE measurements were possible in 97% of longitudinal strain and 100% of radial strain and circumferential strain. Intra- and inter-observer variability of longitudinal, radial and circumferential strain was good. Longitudinal strain and radial strain were significantly lower in neonates with CoA (-13.98% versus -17.80%, $p = 0.031$, and 19.50% versus 38.35%, $p < 0.0001$, respectively). Circumferential strain was not modified. To predict CoA, a longitudinal strain value of -16.17% provided a sensitivity of 70% for a specificity of 80%. A radial strain value of 28.1% provided a sensitivity of 80% for a specificity of 80%.
Conclusion: LV STE is feasible in newborns with PDA and seems to be efficient for predicting the risk of CoA. Further studies are needed to confirm these results.

P1292 - IMPROVING THE RESPONSE TIME QUALITY AND SAFETY OF STAT X RAYS IN THE CONGENITAL CARDIAC INTENSIVE CARE UNIT

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Background: Arrival of the postoperative congenital heart patient to the congenital cardiac intensive care unit (CCICU) is a critical time in the patient's course. Chest x-rays (CXRs) are effective in quickly identifying issues leading to adverse outcomes. -Our primary goal was to reduce the time to completion of a STAT CXR from a standard of 60 minutes to 15 minutes. -Our secondary goal was to improve the safety and quality of CXRs by ensuring that a nurse was assisting patient positioning and ensuring the gonad shield was used.
Method: -Baseline data was collected for time to stat CXR and quality and safety metrics. -A process for a "super STAT" CXR was developed. Completion time of less than 30 minutes was the initial benchmark. -Bedside nurses and imaging technologists were educated. -Initial data was collected over 4 months. Follow-up data was obtained 6 and 12 months later.
Results: During the initial study period, the average response time to "super STAT" CXR was 18 minutes (range 36-67 minutes) -Follow up data showed the improvement in response time to 12 minutes (range 9-35 minutes) Patient safety metrics improved. -No repeat CXRs were required in the study period, reducing patient radiation exposure. -Gonad shielding was used 100% of the time. -Improvement was sustained during the follow-up period Quality metrics also improved. -Improvement was sustained during the follow-up period.

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Conclusions: Development of a "super STAT" CxR process leads to a more rapid CXR completion time with fewer significant delays. Increased teamwork and collaboration between nursing and imaging technicians led to improved safety. -Gonad shielding was consistently used and documented. -Radiation exposure was reduced as no CXRs needed retaking. Increased teamwork and collaboration between nursing and imaging technicians led to improved quality. -Patient positioning was more appropriate. -Lines and tubes were moved away from the chest. These results were sustained at 12 months.

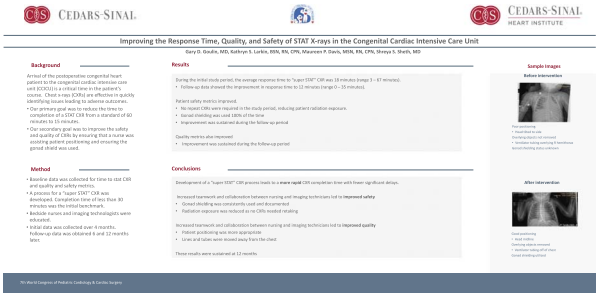

Figure 1.

P1299 - ANALYSIS OF CARDIAC MICROSTRUCTURE AND MYOCYTE AGRREGATES ORGANIZATION IN FETAL HEARTS WITH CONGENITAL HEART DISEASE BY X RAY PHASE CONTRAST SYNCHROTRON IMAGING

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Background: Understanding the complexity of heart morphogenesis and the associated functional consequences of congenital heart disease (CHD) is essential for providing appropriate treatment strategies. Since our knowledge on the microstructure of the whole structurally abnormal heart in fetal & paediatric CHD is limited, novel imaging approaches offered by synchrotron facilities can provide structural detail currently not available otherwise. Our aim is to visualize and quantify cardiac ultrastructure in hearts with altered fetal development using X-ray phase-contrast synchrotron radiation-based micro-CT.
Methods: Two fetal hearts, one normal and one with Tetralogy of Fallot (ToF) of 15 weeks of gestation were selected from the UCL Cardiac Archive. While the specimens were kept in formalin, they were settled in water as supporting medium for acquisition. X-ray phase-contrast synchrotron radiation-based micro-CT was performed at 3.6μm resolution in the Diamond Light Source, I13-2 beamline facilities. Several acquisitions were necessary to cover the whole heart along its long axis. The series were reconstructed using state-of-the-art filtered back projection.
Results: Detailed visualization of different cardiac structures such as aortic and pulmonary valves, papillary muscles, tendinous cords of the mitral valve were obtained. The ventricular septal defect as well as the overriding aorta were clearly visible in the ToF fetal heart. In the normal heart, there is a transition in angle of myocyte aggregates from epicardium to endocardium with circumferential layers within both the LV and RV. In the case with ToF, a deep trabecular layer of the LV appears more compacted but also more heterogeneous.
Conclusions: We managed for the first time to acquire μ-resolution microstructural datasets of whole human fetal hearts, illustrating normality as well as congenital malformations, resolving myofibre detail and providing information on cardiac microstructure without the need of slicing and sample processing.

P1320 - CHANGES IN OVERALL VENTRICULAR MYOCARDIAL ARCHITECTURE IN THE SETTING OF A PORCINE ANIMAL MODEL OF RIGHT VENTRICULAR DILATION

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Background: Chronic pulmonary insufficiency often leads to myocardial dysfunction and heart failure. It is not fully known why secondary hypertrophy cannot fully protect against the increase in wall stress brought about by the increased end-diastolic volume in ventricular dilation. It has been assumed that mural architecture is not deranged in this situation, but we hypothesised that there might be a change in the pattern of orientation of the chains of cardiomyocytes, which would contribute to contractile impairment.
Methods: We created pulmonary valvar insufficiency by transvascular suturing of its leaflets in seven piglets, performing sham operations in seven control animals. Using cardiac magnetic resonance imaging after 12 weeks of recovery, we demostrated significantly increased right ventricular volumes in the test group. After sacrifice, diffusion tensor imaging of their hearts permitted measurement of the orientation of the long chains of aggregated cardiomyocytes.
Results: The helical angles in the right ventricle approached a more circumferential orientation in setting of right ventricular dilation ($p = 0.007$), with an increased proportion of surface-parallel chains. In contrast, this proportion decreased in the left ventricle. Also in the left ventricle a higher proportion of E3 angles with a value around zero was found, and conversely a lower proportion of angles was found with a numerical higher value. In the dilated right ventricle the proportion of E3 angles around -90 degrees is increased, while the proportion around 90 degrees is decreased.
Conclusion: Contrary to traditional views, there is a change in the orientation of both the left and the right ventricular cardiomyocytes subsequent to right ventricular dilation. This will change their direction of contraction and hinder the achievement of normalization of cardiomyocytic strain, affecting overall contractility. We suggest that the aetiology of the cardiac failure induced by right ventricular dilation may be partly explained by morphological changes in the myocardium itself.

P1322 - POST MORTEM MICRO CT SCANNING OF FETAL HEART

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Congenital heart defects (CHD) represent the most common malformation with approximately 20% of stillbirths and 30% of neonatal deaths due to congenital defects. Fetal cardiac autopsies are crucial in counseling parents after the fetal /child death. Nowadays, the diagnosis of CHD is often anticipated to the first trimester of pregnancy, however the feasibility of routine autopsy in small fetuses is only in about 30% of cases. Conventional MRI or high-field micro-MRI offer only a low resolution in small fetuses and their use in clinical setting is limited by cost, time and space constraints. Therefore, micro CT scan was explored. Micro CT is X-ray imaging in 3D as in hospital CT scans on a smaller scale with massively increased resolution.
Methods: Iodine contrast micro-CT scans were used to examine 22 fetal/neonatal hearts. Images were obtained from routinely fixed 7 whole human fetuses (0.1-90 g, 7-17 w.g.), 14 isolated fetal hearts (0.1-20 g, 11-27 w.g.) and 1 isolated neonatal heart. Samples were scanned using an isotropic resolution of 18 μm and reconstructions were interpreted jointly by four fetal pathologists, fetal cardiologist and radiologist. Cases with gestational ages ≥ 13 w.g. underwent also conventional autopsy with histological sample. Post-mortem assessments were compared to echographic diagnosis.
Results: Micro-CT identified all anatomical structures and abnormalities documented by prenatal echography. In 7 whole fetuses micro-CT excluded structural anomalies. In 14/22 cases it provided all informations obtained with invasive autopsy and in 7 of them it furnished additional diagnostic details. In the neonatal case (object of litigation for a missed diagnosis of CHD) it was useful for discussion among courtroom experts.
Conclusions: Micro-CT study is useful for refining or modifying prenatal echographic diagnosis in small fetuses or neonates. It was also found useful in case of diagnostic controversies.

P1333 - VITAMIN D DEFICIENCY RICKETS AND CARDIOVASCULAR FEATURES IN EARLY CHILDHOOD

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Introduction: -Vitamin D deficiency and hypocalcemia is one of the treatable cardiomyopathy causing cardiac failure in children. This study aimed to correlate vitamin D deficiency with the cardiovascular status in children under 3 years of age.
Methods: -Fifty newly diagnosed cases of vitamin D deficiency rickets under 3 years with atleast 2 clinical signs of rickets and Vitamin D levels (<20 ng/ml) were studied for their cardiovascular status. Children with Congenital heart disease and acquired heart diseases were excluded. Serum calcium/phosphorous/alkaline phosphatase levels, 25 (OH) Vitamin D levels, ECG and 2D Echocardiography were done in all. All the ECG and ECHO parameters were compared with age related norms and body surface area (z score).
Results: - Mean age of children was 11 months with 24/50 (48%) between 3-6 months. The male to female ratio was 1.9:1. 20/50 (40%) were exclusively breastfed, 8(16%) formula fed, 15(30%) were on milk based diet and 7(14%) were on normal diet. 36/50 (72%) had low total serum calcium levels. Prolonged QTc interval (12%) and left axis deviation were the common ECG abnormality noted. 2 D Echocardiogram showed 11(22%) had LVH, 10 (20%) had LV systolic dysfunction, 9 (18%) had LV dilation, 8 (16%) had diastolic dysfunction,7 (14%) had MPI >0.38, and 4 (8%) presented with dilated cardiomyopathy. Abnormal z scores were noted with Vitamin D deficient patients with a statistically significant negative correlation between LVIDd and total calcium levels. Spearman's rank correlation coefficient between vitamin D levels and various LV dimensions was statistically insignificant.
Conclusion: -Left ventricular dysfunction with dilatation and hypertrophy are noted in children with vitamin D deficiency though they are asymptomatic . Hence Cardiac screening should be a standard of care in all children with vitamin D deficiency as the cardiac dysfunction is reversible on supplementation.

P1335 - CARDIOVASCULAR MANIFESTATIONS OF CHILDHOOD ONSET HYPOTHYRIODISM

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Introduction: Thyroid hormones exert a direct cellular effect on almost all the tissues of the body including heart. Limited studies are available in the country to assess the various cardiovascular parameters in hypothyroid children and the impact of treatment on these parameters.This study is aimed at studying the cardiac dysfunction in hypothyroidism by electocardiography (ECG) and echocardiography (2DECHO).
Methods: 32 newly diagnosed hypothyroid pediatric patients were studied over a period of 18 months. They were clinically evaluated and underwent relevant investigations, including thyroid profile estimation, cardiac evaluation using E.C.G. and 2D – Echocardiography /colour Doppler.29 patients were reviewed after 3 months of treatment. A p-value of <0.05 was considered statistically significant.
Results: 18/32 (56.3%) were above 10 years with female preponderance (68.8%). Mean value of TSH and Free T4 were 205.58 ± 267.33 μIU/ml and 0.67 ± 0.37 ng/dl respectively. Symptoms and signs were common with TSH (thyroid stimulating hormone) above 50μgm/ml and was statistically significant. Dyslipidemia was found in 22 (68.75%) cases. The most common ECG abnormality observed was prolonged PR interval in 14 (43.8%)cases. The most common echocardiographic finding was septal hypertrophy, seen in 16(50%) cases, followed by left ventricular posterior wall hypertrophy (LVPW) (43.8%), LV dilatation (25%), pericardial effusion (25%) and cardiac dysfunction in 4 cases. On 3 months follow up of 29 subjects, significant improvement of all the deranged parameters were noted with achievement of euthyroidism.
Conclusion: Children with hypothyroidism have significant cardiovascular abnormalities which are proportionally related to TSH level and these changes are completely reversible with treatment. Hence, early diagnosis with optimal treatment reduces the cardiac morbidity in these children.

P1337 - PULMONARY HYPERTENSION IN CHILDREN WITH CONGENITAL HEART DIEASE

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Aim: To note clinical profile of children with congenital heart disease (CHD) with an increased pulmonary blood flow and correlate the grade of Panama functional class (2011) with Echocardiographic data.
Method: 50 children with CHD with an increased pulmonary blood flow based on 2 D Echocardiogram were compared with 50 age and sex matched controls . Demographic data, clinical profile, chest x-ray , echocardiographic data, treatment and severity of Pulmonary hypertension (PAH) as per Panama class was noted .
Results: 40/50 (80%) presented in their first year of life. The most common presenting symptom was dysponea (80%). Tachycardia and tachypnoea were statistically significant in the study group than the controls (p value <0.01). Stunting, wasting and underweight on WHO growth charts was significant (p value for both underweight and wasting <0.01 and for stunting 0.025). 81.5% of children had thrill with PAH and significant association was also noted with palpable P2 and PAH . 25/50 (50%) belonged to class 3 B of Panama functional class and all Class 3B cases had severe degree of malnutrition. The severity of PAH in class 3B, class 3 A and 2 are 52.6%, 42.1% and 5.3% respectively. On comparing degree of PAH on Echocardiogram with Panama class no statistically significant correlation found (p value 0.741). Children with PAH had statistically significant IVS , RV free wall thickness and RV FAC grade as compared to controls.Surgery was advised in all cases of PAH and this was statistically significant (p value 0.01).
Conclusion: Children with CHD and an increased Pulmonary blood flow present with PAH early and are usually of grade 2 severity at presentation. However no linear association was noted between severity on clinical classification and degree of PAH on Echocardiography.

P1355 - FETAL HYPERTROPHIC CARDIOMYOPATHY AS AN EARLY MARKER OF MATERNAL HYPERGLYCEMIA

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Background: Gestational diabetes mellitus (GDM) cause maternal hyperglycemia, which may lead to fetal hypertrophic cardiomyopathy (HCM), and cardiac diastolic function impairment. The aim of the present study was to assess the prevalence of HCM in fetuses of pregnant women with GDM before their treatment using fetal echocardiography.
Material and Methods: This cross-sectional study included 63 fetuses of mothers with GDM and was conducted before initiating treatment. It was conducted in a public maternity clinic at southern Brazil. according to the local regulations for good clinical practices, specifically those of the National Resolution of the National Health Council (CNS, 466/12), after approval from the Research Ethics Committee (CEP011/12) of the institution. It included singleton fetuses of pregnant women diagnosed with GDM, without malformations and other diseases. GDM diagnosis was based on the criteria provided by the American Diabetes Association and an ultrasound was performed between the 24th and 28th gestational weeks. All variables were descriptively analyzed, with quantitative variables expressed as means and standard deviations. Data were statistically analyzed using the Statistical Package for the Social Sciences software, version 21.0.
Results: HCM was confirmed in 54% of the fetuses. Fetal abdominal circumference (FAC) was normal in 73.01% fetuses. HCM was also observed in 50% of fetuses with normal FAC, which reflects the occurrence of interventricular septum alterations prior to FAC alterations.
Conclusions: The result suggests that the HCM reflects the maternal hyperglycemia before FAC alterations. Therefore, the high prevalence and early occurrence of HCM in this population suggest that this is one of the first effects of maternal diabetes on fetuses.

P1356 - CARDIAC FUNCTION OF NEWBORN BABIES OF PREGNANT PATIENTS WITH GESTACIONAL DIABETES MELLITUS

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Background: Gestacional diabetes mellitus (GDM) promotes structural and functional abnormalities in the newborn heart. In this context our goal is to evaluate the cardiac function of newborn babies of pregnant patients with GDM by means of Doppler echocardiography data.
Materials and Methods: Were analysed 48 echocardiography exams from newborns's GMD mothers with good clinical control durant pregnancy and who performed routine echocardiographic evaluation during the neonatal period. In these babies the cardiac variables analyzed were: ventricular septum thickness and posterior wall of the left ventricle by M mode, global cardiac funtion by myocardial performance índex (MPI, also known as Tei índex), diastolic function by reason E/A of both mitral and tricuspid valve and sistolic function by the shortening fraction measurement of left ventricle. It was conducted in a private maternity clinic at southern Brazil according to the local regulations for good clinical practices, specifically those of the National Resolution of the National Health Council (CNS, 466/12), after approval from the Research Ethics Committee (CEP 1.572.265) of the institution.
Results: The newborns presented in average 18 ± 12 days old and the incidence of myocardial hypertrophy was 6%. About the echocardiography variables of heart, the shortening fraction was abnormal in 7% and diastolic function of left and right ventricle was irregular with 47% e 30%, respectively. TEI índex was abnormal in 51% of the right ventricle and 57% of the left ventricle.
Conclusions: The newborn's GMD mothers with good clinical control presented low índex of hypertrofic cardiomyopathy at birth and even in the absence of cardiomyopathy the cardiac function has been abnormal in some parameters.

P1374 - REFERRAL PATTERN AND SPECTRUM OF CONGENITAL HEART DISEASES DIAGNOSED AT FETAL ECHOCARDIOGRAPHY IN EASTERN INDIA

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Background and Aims: Fetal echocardiography is a potential but inadequately utilised tool to detect and prevent major congenital heart diseases (CHD). Our study was designed to find out the referral pattern and spectrum of CHD diagnosed during fetal echocardiography in Eastern part of India.

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Materials and Methods: A retrospective analysis of all fetal echocardiography performed over two years period (August 2014 to July 2016) in our centres of Kolkata was done. The primary indications for referral for fetal echocardiography were obtained from the obstetric referral prescriptions. Patient details and outcome data were collected from fetal echocardiography report. Fetal echocardiography was performed jointly by same pediatric cardiologist and fetal medicine specialist (author and coauthor).
Results: Total 772 fetal echocardiography reports performed during the study period were reviewed. Gestational age varied from 17 weeks to 37 weeks (Median 22 weeks). 54 women had abnormal fetal cardiac findings (58 studies, 7.5% of total studies). The most common referral for fetal echocardiography was related to maternal indications (283/772). Other indications were abnormal findings in antenatal ultrasonography (186/272), family history of CHD (35/772), prenatal routine screening (236/772), and IVF (32/772). The highest yield of significant abnormal findings was found among patients referred with abnormal prenatal ultrasonography (39%). Whereas detection rate for unselected routine screening was less than 1% (2/236). We analysed the pattern of CHD detected in abnormal scans. Most common finding was conotruncal abnormalities (17/54) followed by single ventricle physiology (15/54). Others included 8 VSD, 4 AVSD, 4 significant arrhythmias, 2 Ebstein's anomaly and 4 had other cardiac abnormalities (tumor/pericardial effusion). Insignificant arrhythmias were excluded.
Conclusions: Majority of referral for fetal echocardiography were for abnormal prenatal ultrasonography, usually during anatomy scan between 18-20 weeks. Routine fetal echocardiography for unselected population has very low detection rate.

P1389 - MEDIUM TERM OUTCOME OF PRENATALLY DIAGNOSED HYPOPLASTIC LEFT HEART SYNDROME AND IMPACT OF A RESTRICTIVE ATRIAL SEPTUM DIAGNOSED IN UTERO

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Background/Hypothesis: Surgical outcome data does not reflect outcome of prenatally diagnosed fetuses with hypoplastic left heart syndrome (HLHS); historical data has shown 46.9% overall survival of the intention-to-treat group (ITT). Our aim was to describe outcome of prenatally diagnosed fetuses in the current era.
Methods: Retrospective review of prenatally diagnosed HLHS at Birmingham Women's Hospital over a 10-year period, estimated due dates 01/8/2006 to 31/07/2016. HLHS variants and ventricular disproportion were excluded. Outcome and prenatal variables (Table) were investigated.
Results: We identified 160 fetuses; outcome information was available for 157. There were 44 (28.0%) terminations of pregnancy, 3 (1.9%) intrauterine deaths, 7 (4.5%) babies had compassionate care. There was ITT in the remaining 103/157 (65.6%). Of the ITT group, there were 4 deaths before intervention (3.9%), 99 (96.1%) underwent intervention: a Norwood procedure was performed in 92 (92.9%), an initial hybrid in 6 (6.1%), 1 had coarctation stenting. Survival from birth at 6-months, 1-year and 5-years respectively was 90.3%, 83.5%, 72.5%. Altogether 65 patients are currently alive: 5 with a Norwood circulation, 35 with a cavopulmonary shunt and 24 Fontan circulation (one had cardiac transplantation). Overall survival in the prenatal cohort was 65/157 (41.4%). Multivariable analysis shows a restrictive atrial septum (RAS) is an important factor associated with death, after variable selection HR 1.98 (95% CI 0.93-4.19). There were 6 survivors in this group (n = 15):2 have not completed the Fontan pathway due to unfavorable haemodynamics, 3 are post cavopulmonary anastomosis (all taking sildenafil) and 1 has completed Fontan circulation.
Conclusion: Medium-term outcomes of prenatally diagnosed HLHS have improved however there remains a significant mortality particularly in fetuses with in-utero diagnosis of a RAS where completion of Fontan is particularly unlikely.

Table.

Factor
Restrictive atrial septum
Extra-cardiac anomalies
Tricuspid valve regurgitation
Endocardial fibro-elastosis
Aortic atresia/mitral atresia
Hydrops/pericardial effusion
Estimated fetal weight third trimester

P1397-3D MOTION CORRECTED FETAL CARDIAC MRI EXPERIMENTAL VALIDATION AND CLINICAL APPLICATION

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Background: The accurate diagnosis of congenital heart disease in the fetus is crucial to provide relevant counseling and plan postnatal care. However, visualisation of the extracardiac vasculature is often challenging using echocardiography alone, and MRI, whilst safe, is highly limited by uncontrolled fetal motion.
Materials and Methods: We sought to validate the use of a novel motion-correction algorithm by first applying it to an artificially motion-corrupted 3D balanced steady-state free precession (3D-SSFP) volume, acquired from a one-day old neonate with coarctation of the aorta under general anesthetic with cardiac and respiratory gating. Nine non-contiguous stacks of single images were sythesised (equivalent to standard fetal data), which were then processed by the motion-correction algorithm to produce a three-dimensional volume which could be compared to the original 3D-SSFP sequence. Following this initial experiment, the same technique was applied directly to fetal cases using multiple T2-weighted single-shot fast spin echo sequences, across a range of gestational ages and diagnoses.
Results: The postnatal motion corrected volume generated was qualitatively identical to the "ground-truth" 3D-SFFP volume, demonstrating the same anatomical features (figure 1). To date we have applied this technique to twenty-eight fetuses between 24-36 weeks gestation (mean 32 weeks), predominantly cases with aortic arch abnormalities (coarcation of the aorta, potential vascular rings) and those with undetermined pulmonary arterial or venous anatomy (figure 2). In each case to date, postnatal imaging and/or

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surgery has confirmed antenatal findings. In some cases, antenatal imaging has provided additional useful insights to inform postnatal management (figure 3).

Conclusions: We have used an artificially motion-corrupted neonatal 3D-SSFP volume, acquired under general anaesthetic with cardiorespiratory gating, to validate a novel motion-correction algorithm for use in fetal cases with promising early results. This technique offers the potential to be a powerful adjunct to fetal echocardiography in cases where the vascular diagnosis would otherwise not be fully resolved before birth.

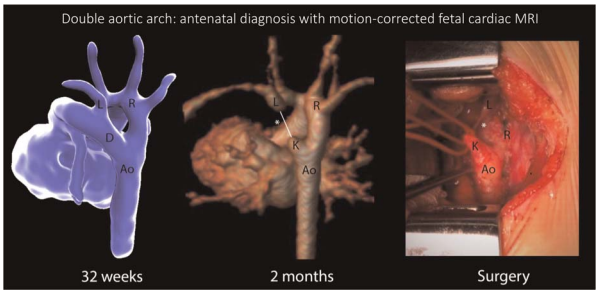

Figure 1.

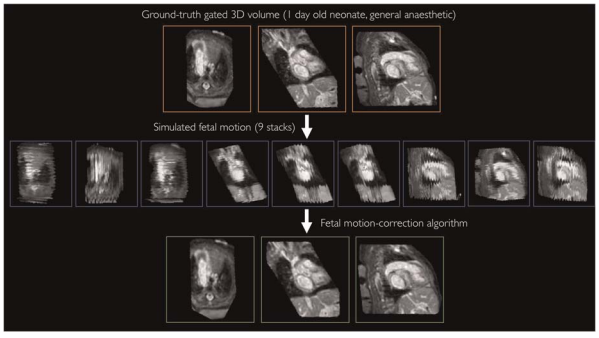

Figure 2.

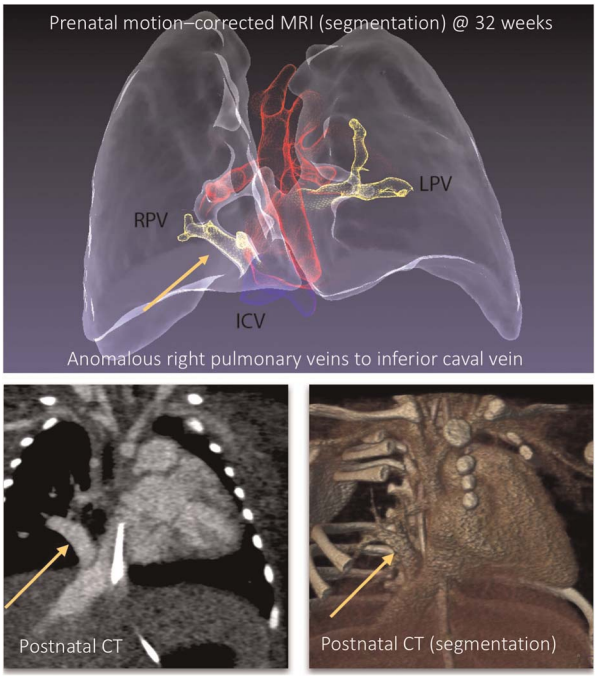

Figure 3.

P1399 - RESTING AND EXERCISE ARTERIAL MECHANICS IN ANTHRACYLINE TREATED LONG TERM SURVIVORS OF CHILDHOOD CANCERS

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Background: We aimed to determine arterial mechanics at rest and during exercise in anthracycline-treated long-term survivors of childhood cancers.

Methods: Eighty-seven (50 males) anthracycline-treated long-term survivors of childhood cancers aged 24.1 ± 5.3 years were studied at 15.1 ± 5.6 years after completion of treatment and 60 (30 males) age-matched healthy subjects were recruited as controls. The blood pressures in the right arm were measured by oscillometry, while radial augmentation index (rAI) was determined by applanation tonometry. Carotid arterial stiffness and intima-media thickness (IMT) were assessed using high-resolution ultrasound. Arterial stiffness and blood pressures were determined both at rest and during submaximal supine bicycle exercise testing with achievement of a target heart rate of 70% of age-predicted maximum.

Result: At rest, patients had significantly greater carotid IMT (0.44 ± 0.15 mm vs 0.41 ± 0.01 mm, $p < 0.001$), carotid arterial stiffness (4.04 ± 0.68 vs 3.85 ± 0.61, $p < 0.001$), rAI (64 ± 15% vs 57 ± 13%, $p = 0.008$), and rAI adjusted for a heart rate of 75/min (63 ± 14% vs 58 ± 12%, $p = 0.020$) than controls. During submaximal exercise, patients had significantly greater increase in carotid stiffness than controls ($p < 0.001$), while systolic and diastolic blood pressures remained similar between the two groups (all $p > 0.05$). For the whole cohort, the magnitude of exercise-induced increase in carotid stiffness correlated positively with rAI ($r = 0.18$, $p = 0.027$), adjusted rAI ($r = 0.22$, $p = 0.009$), and carotid IMT ($r = 0.32$, $p < 0.001$) and age ($r = 0.21$, $p = 0.011$). In patients, the cumulative anthracycline dose received was found to correlate positively with rAI ($r = 0.24$, $p = 0.027$) and heart-rate adjusted rAI ($r = 0.25$, $p = 0.019$). Furthermore, the magnitude of exercise-induced increase in carotid arterial stiffness was found to correlate with years after completion of anthracycline ($r = 0.23$, $p = 0.031$), but not cumulative anthracycline dose ($p > 0.05$).

Conclusion: Arterial mechanics are impaired at rest and worsened during exercise in anthracycline-treated long-term survivors of childhood cancers, which may adversely increase left ventricular afterload.

P1406 - LOOKING INTO THE HEARTS OF THALASSEMIC CHILDREN

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Objectives: To note spectrum of cardiac abnormalities in transfusion dependent β-thalassemia children by clinical and Echocardiographic evaluation and to correlate its severity with serum ferritin levels.

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Design: Observational study at an Urban tertiary referral children's Hospital.
Methods: 100 thalassemic children who received more than 50 blood transfusions and had serum ferritin level >2500 ng/ml were included. Detailed medical history, anthropometry, transfusion details, examination including cardiovascular system were noted. Echocardiogram, Colour Doppler and Tissue Doppler Imaging (TDI) were done to assess cardiac functions. Cardiac dimensions, functions, mass and index were assessed by standard Echocardiographic techniques and were compared to paediatric norms. TDI was performed to assess myocardial involvement.
Results: Cardiac symptoms were largely subclinical. 13 children had tachycardia and 10 were hypertensive for their age. 8/100 had left ventricular dilatation. 85 children had diastolic dysfunction which was seen more in children with Hb <8 g/dL, increasing years of transfusion therapy and increasing ferritin levels (p value: 0.042). 13/100 children had systolic dysfunction. There was a significant correlation between age and systolic dysfunction (p value: 0.017). 32 children had a high cardiac output. There was a significant correlation between increasing serum ferritin levels and TDI abnormalities (p value: 0.006). 45 children had right ventricular dysfunction. There was significant correlation between age and right ventricle dysfunction (p value 0.028).
Conclusion: Cardiovascular manifestations remain predominantly silent in thalassemic children. On Echocardiographic evaluation, LV dilatation, LV diastolic and systolic dysfunctions, impaired myocardial performance, increased cardiac output, moderate TR and RV dysfunction were noted. LV diastolic dysfunction was more common than systolic dysfunction and was noted more in children with longer duration of transfusion therapy and high serum ferritin levels. TDI complimented the conventional Echocardiography in diagnosing diastolic dysfunction.

P1408 - DIFFUSE PULMONARY ARTERIOVENOUS MALFORMATION IN CHILDREN ESSENTIAL VALUE OF CONTRAST ECHOCARDIOGRAPHY

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Background: Pulmonary arteriovenous malformations (PAVMs) although rare, is an important consideration in cyanotic patients of unknown cause. We report 3 diffuse PAVM cases in children detected by contrast echocardiogram.
Case 1: A 1-year-old boy with recurrent respiratory infections since 1-month-old with incidental finding of dextrocardia and situs inversus. He presented at 10-month-old with 2 weeks history of fever and cough with associated respiratory distress requiring ventilator support. He was positive for pulmonary tuberculosis and treatment started. His oxygen saturations remain in low 80 s despite of his clinical improvement. He was cyanosed and clubbed. Echocardiography showed dextrocardia, normal cardiac structure with positive bubble study which raised the suspicion of PAVM. A high resolution computed tomography (HRCT) of the thorax showed no obvious intra-pulmonary shunting or bronchiectatic changes. Pulmonary angiogram confirmed diffuse PAVM.
Case 2: 11-year-old girl with history of right cerebral abscess undergone a burrhole and aspiration. 3 years later she had recurrent abscess at left temporoparietal region, had surgical drainage and 6 weeks of intravenous antibiotics. She had central cyanosis, clubbing, oxygen saturation 85% and upper motor neuron lesion signs. Chest x-ray, CT pulmonary angiogram (CTPA) were normal and echocardiography showed a normal cardiac structure with positive bubble study. Pulmonary angiogram confirmed diffuse PAVM.
Case 3: A 4-year-old boy with history of increasing cyanosis and reduced effort tolerance.Both respiratory and cardiovascular examination were unremarkable. CTPA was normal with positive bubble study on echocardiography. Pulmonary angiogram showed diffuse PAVMs. There were no pulmonary hypertension in all patients.
Conclusion: PAVM is an important diagnosis to consider in persistent hypoxaemia in the absence of respiratory and cardiac disease. Normal HRCT thorax findings does not rule out diffuse PAVM. Hence, contrast echocardiography is essential in diagnosing PAVM. Pulmonary angiogram should be reserved for therapeutic purposes.

P1414 - FETAL ECHOCARDIOGRAPHY FOR RISKY PREGNANCIES IN SULAIMANI GOVERNMENT KURDISTAN IRAQ

Aso Salih[1]
Sulaimani University, Pediatrics, Sulaimanyah-Iraq Diagnosis of congenital heart disease during fetal life not only identifies the cardiac lesion but may also lead to detection of associated abnormalities. This information allows a detailed discussion of the prognosis with parents

Objective: The aim of this study is to evaluate our experience and results of fetal echo for risky pregnancies in sulaimany.
Method: This is prospective descriptive study conducted on 450 pregnant ladies referred for fetal echocardiography from 1st January 2013 still ongoing. Echocardiography done at their first visit at 16-22 weeks in 330 ladies, at 22-28 weeks in 70 cases and 31weeks and above in 50 ladies. Also 2nd visit echo done for 60 patients for follow up at 31-33 weeks gestation.
Result: Among 450 pregnant ladies seen, 98 cases found to have findings, of which 27 diagnosed as hypertrophic cardiomyopathy (just one persist after 6 months post labour), 25 cases as ASD (just 10 left with the defect after labour), 14 diagnosed as a case of tricuspid atersia and single ventricle physiology, 5 AVSD, 5 arrhythmia with hydropis, 5 with MV and TV abnormalities, 4 AS, 3 hypoplastic left heart syndrome, 3 PS, 2 l-TGA and one Coa . For low risk populations of defects no action undertaken, for high risk population decision for termination is one of options Fetal echocardiography examination specifity was 97.8% and sensitivity was 98.3%, positive predictive value was 93.8%, negative predictive value was 99.4%.
Conclusion: The present study showed that fetal echo has great value in detection of congenital heart diseaseprognosis and outcome in high risk pregnancies. It's recommended to familiarize our obstetrician about this important diagnostic test. We should train our ultrasonographer and pediatric cardiologist to increase awareness about this diagnostic.

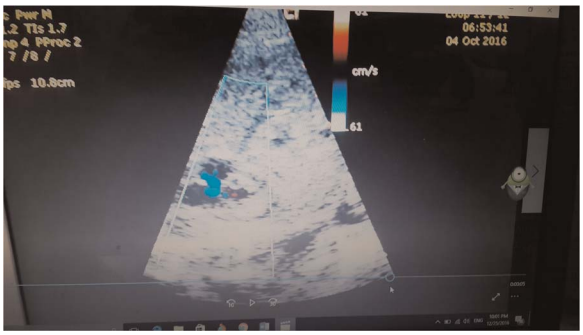

Figure 1.

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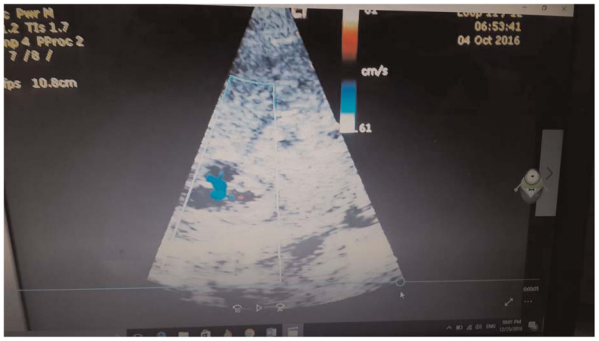

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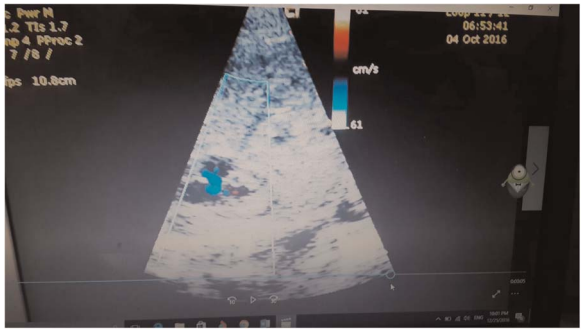

Figure 3.

P1429 - ACCURACY OF COMPUTED TOMOGRAPHY FOR DETECTING GREAT VESSEL STENOSIS OR HYPOPLASIA IN CONGENITAL HEART DISEASE PRIOR TO SUPERIOR BIDIRECTIONAL CAVOPULMONARY CONNECTION COMPARISON WITH CARDIAC CATHETERIZATION AND SURGICAL FINDINGS

Laura Vazquez-Garcia[1], *Monther Obeidat*[1], *Giovanni Di Salvo*[1], *Olivier Ghez*[1], *Guido Michielon*[1], *Michael Rubens*[2], *Zdenek Slavik*[1], *Michael L Rigby*[1], *Alain Fraisse*[1], *Sylvia Krupickova*[1]
Royal Brompton Hospital, Paediatric Cardiology, London-United Kingdom[1]*; Royal Brompton Hospital, Radiology, London-United Kingdom*[2]

Background: This study sought to investigate diagnostic accuracy and safety of computed tomography (CT) in assessing great vessels stenosis/hypoplasia prior to superior bidirectional cavopulmonary connection (BCPC) compared with cardiac catheterisation (CC) and surgical findings.
Methods: Twenty-seven patients (37% after Norwood operation) underwent CT prior to BCPC (16 patients out of them also CC) between January 2010 and June 2016. At the time of BCPC, median age was 229days and median weight 8.3 kg. Proximal and distal pulmonary artery branches, aortic isthmus and abdominal aorta were measured.
Results: Nine right and 11 left pulmonary artery branches were enlarged at the time of surgery. CT did not miss any stenosis/hypoplasia of pulmonary arteries and reported 2 cases of pulmonary arteries hypoplasia that were subsequently considered to be normal at the time of BCPC. The accuracy of CT for detecting stenosis/hypoplasia of either pulmonary artery was 99.7%. No coarctation of aorta was noted at CT and thus not formally assessed at the time of surgery. In terms of absolute vessel measurements and their Z-scores, there was excellent agreement ($r = 0.98$ for both) between CT and CC, very low bias (0.71mm, 0.48; respectively), and clinically acceptable limits of agreement (−0.9 to +2.3mm; -0.7 to +1.7, respectively). Patients received significantly higher radiation dose at CC than CT (median 2.7mSv, IQR 1.7,3.6 versus median 1.2mSv, IQR 1.0,1.9, respectively; $p < 0.040$). Four patients (25%) suffered minor complications from CC and there were no complications from CT scan. All cardiac catheterisations were performed under general anaesthesia whereas no sedation was required for any of the patients for CT.
Conclusions: CT may replace CC for identification of great vessel stenosis or hypoplasia in selected patients before BCPC when there is no suspicion of requiring any intervention prior to surgery. CT requires less radiation, carries less morbidity and can be performed without sedation.

P1464 - PATENT DUCTUS ARTERIOSUS MORPHOLOGY IN DUCTAL DEPENDENT PULMONARY CIRCULATION – HOW DOES COMPUTED TOMOGRAM ANGIOGRAPHY HELPS

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Background: Patent Ductus Arteriosus (PDA) in duct dependent pulmonary circulation(DDPC) has a diverse morphology. The objectives of this study are to assess the role of computed tomogram angiography(CTA) to classify type of PDA in DDPC, identify pattern of PDA morphology in different type of ventricular morphology (biventricular, univentricular and Pulmonary atresia with intact septum) and evaluate the impact of PDA morphology in the suitability for PDA stenting.
Methods: This prospective study, performed from January 2013 to December 2015, in all patients with DDPC using a 64 slice dual source CT scanner. The PDA was classified based on its site of origin from the aortic arch either from descending aorta-type I, distal arch-type II, proximal arch-type III or from subclavian artery-type IV.
Results: Total 114 patients, age between 2 days to 6 months and weight from 2 kg to 6.9 kg recruited. Seventy nine patients (69%) were on I.V Prostaglandin infusion. 13(11.0%) patients had type I, 74(65.0%) patients type II, 16(14.0%) patients had type III and 9 (8.0%) patients had type IV PDA. Tortuous PDA was found in 37 (32.4%) majority in those with type II and III PDAs. Site of insertion was onto the left pulmonary artery(LPA) in 62(54.4%), right pulmonary artery(RPA) in 20(17.85%) and main pulmonary artery(MPA) in 30(26.78%). Ipsilateral branch PA stenosis found in 75(66.9%) of patient. The majority of PAIVS (92.7%) has type I PDA while in those with biventricular and univentricular lesions have mainly type II or III PDA. Type IV PDA are more common in biventricular lesion. PDA stenting was successful in 69.2% in type I, 41.9% in type II, 37.5% type III and 67.7% type IV.
Conclusion: CTA provides detail delineation of PDA morphology in DDPC lesions. Tortuous type II and III PDA are less likely to be amenable for PDA stenting.

P1469 - ACCURACY OF ECHOCARDIOGRAPHY IN THE DIAGNOSIS OF TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION

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Hypothesis: Total anomalous pulmonary venous connection (TAPVC) is diagnosed by echocardiography and may be

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confirmed by other diagnostic modalities. The objective of this study was to determine the accuracy of echocardiography in the diagnosis of different types of isolated TAPVC and TAPVC with other congenital heart defects (CHD) and compare it with CT angiography or observation during surgery.
Patients and Method: Data from the medical records of patients 1 day to 15 years of age, from Jan 2010 to Aug 2016 with echocardiographic diagnosis of TAPVC and had confirmation with either at surgery or with CT angiogram was obtained. Accuracy of echocardiography was defined as complete, partial and missed diagnosis and was compared with CT angiogram or surgery.
Results: Total of 47 patients were enrolled, 33 (70.2%) were males, and 14 (29.8%) were females. 41 (87%) were infants, 18 (38%) were less than 1 month and 6 (13%) were between 1-15 years. CT angiogram was done in 30 patients. 32 (68%) had isolated TAPVC, while 15 (32%) had associated CHD. 18 (38%) were supracardiac, 17 (36%) were cardiac, 8 (17%) infracardiac and 4(9%) were mixed type of TAPVC. Echocardiographic diagnosis of TAPVC was complete in 33(66%), partial in 6(13%), missed in 10(21%) of patients. In Isolated TAPVC 27 (84.4%) patients were diagnosed completely and 5(15.6%) patients were missed, while in TAPVC associated with CHD 4 (27%) patients were diagnosed completely, 6 (40%) were diagnosed partially, and in 5(33%) the diagnosis was missed. The overall accuracy of diagnosis by echocardiography had 66% (sensitivity) with 100% positive predictive value. In isolated TAPVC, sensitivity was found to be 84% as compared to TAPVC associated with CHD (27%).
Conclusion: We conclude that the mixed variety of isolated TAPVC and TAPVC associated with complex CHD should undergo CT angiogram for accurate diagnosis.

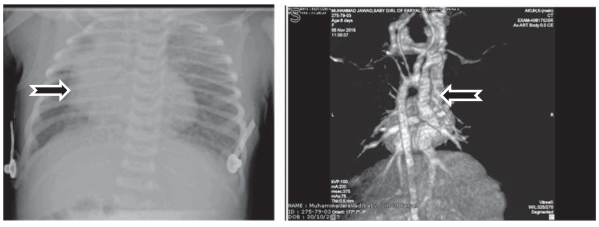

Figure 1.
Case 1: Large pericardial rhabdomyoma (arrow)

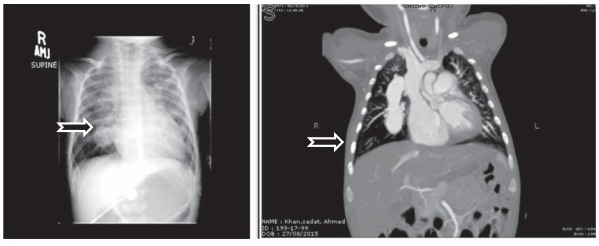

Figure 2.
Case 2: Pericardial rhabdomyoma near left ventricle (arrow)

P1470 - ASSESSMENT OF CARDIAC FUNCTION IN FETUSES OF GESTATIONAL DIABETIC MOTHERS DURING IN THE SECOND TRIMESTER

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Introduction: Fetuses of diabetic mothers may have structural or functional cardiac abnormalities which increase morbidity and mortality. Isolated functional abnormalities have been identified in the third trimester. The aim of the present study was to assess fetal cardiac function (systolic, diastolic and global myocardial performance) in the second trimester in mothers with gestational diabetes, and also to relate it with glycemic control.
Methods: Mothers with gestational diabetes mellitus referred for fetal echocardiography in the second trimester from March 2015 to February 2016 were enrolled as case subjects. Exclusion criteria were pregestational diabetes, presence of structural heart disease, twin pregnancies, intrauterine growth retardation (defined as weight for gestation less than 5th percentile) and polyhydramnios. Non diabetic mothers who had a fetal echocardiogram done between 19-24 weeks for other indications were enrolled as controls. Glycemic control was classified as good if Hb A1c was <6 gm/dl or glucose tolerance test showed serum glucose levels of <160 mg/dl.
Results: 128 mothers were enrolled, 64 were in gestational diabetic group and 64 were in non-diabetic control group. Mean age of mothers was 31 ± 4.9 years (median 29 years) and the mean gestational age at the time of first examination was 23 ± 3.4 weeks (median 22 weeks). Functional variables showed that isovolumetric relaxation and contraction times were significantly prolonged in fetuses of diabetic mothers ($p = 0.001$ and 0.01 respectively). Myocardial performance index and mitral E/A ratios were significantly abnormal in the diabetic group ($p < 0.001$ and $p = 0.03$ respectively)). Mitral annular plane systolic excursion (MAPSE) was less in the diabetic group ($p = 0.01$). The only functional cardiac variable found abnormal in mothers with poor glycemic control was prolonged isovolumetric relaxation time ($p = 0\text{-}03$).
Conclusion: Functional cardiac abnormalities can be detected in the second trimester in fetuses of gestational diabetes and timely intervention can improve postnatal outcomes.

P1471 - PRENATALLY DIAGNOSED LARGE INTRAPERICARDIAL RHABDOMYOMA WITHOUT HEMODYNAMIC COMPROMISE

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Aga Khanuniversity Hospital, Pediatrics, Karachi-Pakistan

Introduction: Rhabdomyoma of the heart is the most common cardiac tumor. It is associated with tuberous sclerosis in 60-80% of cases. In a review of 266 fetuses with cardiac rhabdomyoma, only one had pericardial rhabdomyoma. We present two fetuses with pericardial rhabdomyoma diagnosed prenatally without hemodynamic compromise.
Case 1: 28 year old woman with a gestational age of 32 weeks was referred for fetal echocardiogram which showed a huge intrapericardial tumor measuring 33mm x 18mm with trace pericardial effusion (figure 1). There was no evidence of hydrops fetalis.
Post natal echo showed a large extracardiac tumor intrapericardial with a few hyperechoeic tumors masses in the left ventricular myocardium and in the interventricular septum. None of the tumor masses were producing hemodynamic abnormalities. Conservative management with echocardiographic follow up showed regression in the intrapericardial tumor by one year of age. Magnetic resonance imaging of the brain showed tubers in the brain.
Case 2: 21 year old woman was referred for fetal echocardiography at 26 weeks of gestation with the diagnosis of an extracardiac tumor. There was no hydrops fetalis. Fetal echocardiogram showed a mass measuring 28×20mm in the pericardium adjacent to the left ventricle with no evidence of cardiac compression (figure 2). No intracardiac or intramural myocardial echogenic densities were seen. The diagnosis was intrapericardial rhabdomyoma since the appearance was homogenous as opposed to

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teratoma which has an inhomogeneous appearance. Postnatal follow up showed a reduction in size of the pericardial rhabdomyoma to 14 × 14mm at seven months after birth. The child was referred to a neurologist for screening of tuberous sclerosis.
Conclusion: Primary cardiac tumors are rare and intrapericardial rhabdomyomas are uncommon. Teratoma is the most common intrapericardial tumors, and is easily diagnosed due to the presence of calcified foci and cystic areas. Rhabdomyomas are homogenous.

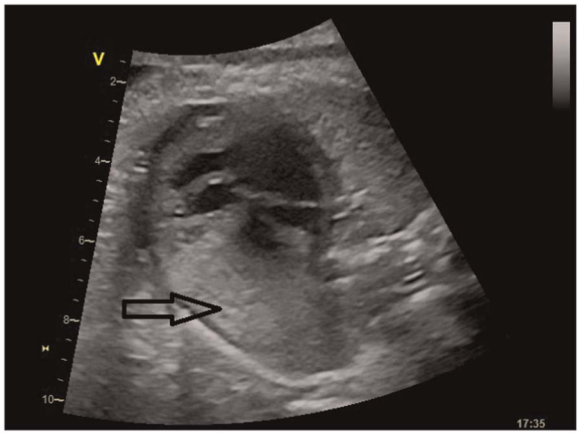

Figure 1.
Case 1: Large pericardial rhabdomyoma (arrow)

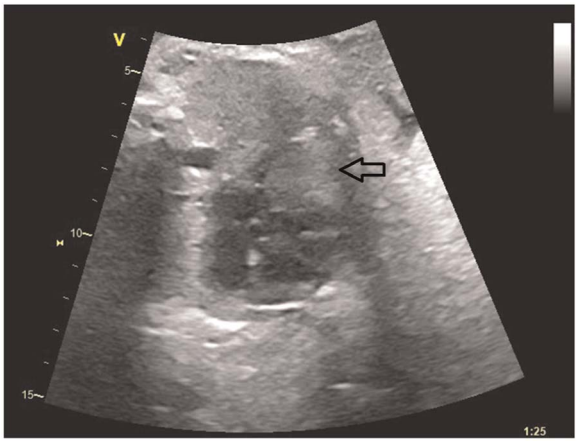

Figure 2.
Case 2: Pericardial rhabdomyoma near left ventricle (arrow)

P1477 - ASSESSMENT OF SINGLE VENTRICLE'S MYOCARDIAL FUNCTION IN GUCH PATIENTS AFTER THE TOTAL CAVOPULMONARY CONNECTION BY TISSUE DOPPLER IMAGING AND MAGNETIC RESONANCE IMAGING

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Objective: Was to estimate the single ventricle's myocardial function in GUCH-patients after the Total Cavopulmonary connection (TCPC) by echocardiography (Echo) and Magnetic Resonance Imaging ().
Material and Methods: From 2003 to 2016 132 patients were performed operation TCPC. Eighteen patients elder 18 y. were included to this work (10 males, 8 female, mean age 21.3 ± 1.9 y, 18-23 years). EDV, ESV, EDI, EF (Simpson's method) recorded by Echo and MRI. Peak S, E, A, ratio E/A, Myocardial Performance Index (MPI) estimated by Echo (TDI).
Results: The peak S (9.2 ± 1.8 mm) by TDI and EF (51 ± 5%) by MRI had direct significant correlation ($r = +0.6$, $p < 0.05$). MPI by TDI (0.51 ± 0.03) and EF (51 ± 5%) by MRI had inverse significant correlation ($r = -0.3$, $p < 0.05$). EDV (58.4 ± 15 ml), ESV (29 ± 8 ml), EF (44 ± 5%) (Simpson method) by Echo did not correlate with the same indexes estimated by MRI (100 ± 18 ml, 46.5 ± 9.1 ml, 51 ± 5% resp.).
Conclusions: Echo indices (peak S, MPI) could use for assessment of single ventricle's myocardial function in GUCH-patients after the TCPC, by their according to the MRI. Volumetric indices and EF assessment by Echo have a poor interdependence with MRI.

P1491 - LEFT VENTRICLE NONCOMPACTATION AND EBSTEIN'S ANOMALY REPORT OF FOUR NEONATAL CASES

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Background: Left ventricular noncompactation (LVNC) is a rare disease characterized by prominent myocardial trabeculations. Clinical features ranges from no symptoms to heart failure. It can be an isolated malformation or associated with other cardiopathies, like Ebstein's anomaly (EA). This association and the clinical behavior in the pediatric literature has rarely been reported. We describe four neonatal cases of LVNC and EA emphasizing on the short and medium term follow-up.
Materials and Methods: This is a retrospective study reviewing patients admitted in the Pediatric Cardiology Unit of the Hospital Sant Joan de Déu diagnosed of EA and LVNC. We collected information on the prenatal and postnatal diagnosis, clinical presentation, management and follow-up.
Results: In the last four years we detected 6 cases of EA with LVNC in 4 cases. The prenatal echocardiography described EA in 2 patients; the other 2 were described as normal although one had increased nuchal translucency. LVNC was detected at birth in 3 patients, and the other one at the first month. Two cases had heart

Table 1.

Patient	Drugs during pregnancy	Prenatal findings	Weight at Birth (grams)	Sex
1	No	EA Septal myocardial hypertrophy	3590	M
2	No	No	2700	F
3	Lithium, Quetiapine, Adalimumab	EA Hypoplastic right ventricle	2810	F
4	No	Increased nuchal translucency	3210	F

Patient	Neonatal echocardiography	Evolution	Interventions	Current age and treatment
1	EA	1 month of age: Acute cardiac failure and Diagnosis of the LVNC	No	4 years. Captopril and carvedilol
2	EA, LVNC, VSD	Neonatal HF and SVT (WPW)	Radiofrequency ablation	3 years. No treatment
3	EA LVNC	SVT	Glenn procedure	2 years. Flecainide
4	EA LVNC	Idiopathic pericardial effusion	No	2 years. ASA

M: male; F: female; EA: Ebstein's Anomaly; LVNC: left ventricle noncompactation; VSD: Ventricular septal defect; HF: Heart failure; SVT: Supraventricular tachycardia; WPW: Wolff-Parkinson-White syndrome; ASA: acetylsalicylic acid

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failure during the first month with satisfactory evolution; however one is still treated with captopril and carvedilol. Two patients had supraventricular tachycardia (SVT). One was treated with flecainide and the other one required radiofrequency ablation due to a Wolff-Parkinson-White syndrome. This patient had a severe EA and required Glenn procedure. One patient receives prophylactic acetylsalicylic acid.
Conclusions: In the last 20 years, we diagnosed 26 cases of EA, but only in the last 4 years we find this rare association. Further, half of the patients had a fetal diagnosis of EA, none of LVNC. That reflects an under pre and postnatal diagnosis. Although an initial adverse clinical outcome was observed in 50% of patients, a satisfactory recovery was seen. Despite these encouraging data, the long-term evolution remains to be clarified.

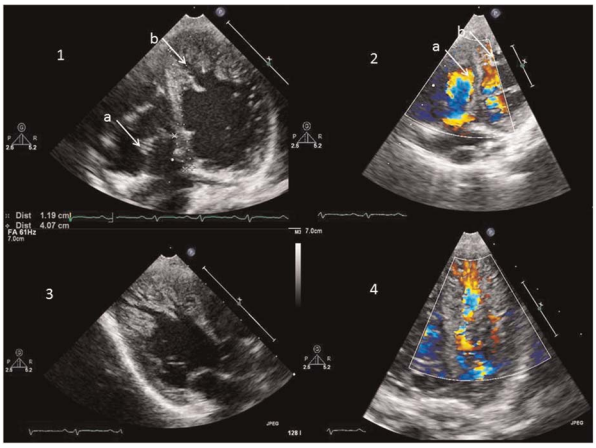

Figure 1.
a) detail of the apical displacement of the tricuspid valve. b) the same image in Color Doppler. 3,4: myocardial trabeculations and deep intertrabecular recesses

P1493 - PREDICTIVE ROLE OF GLOBAL LONGITUDINAL STRAIN IN FUNCTIONAL SINGLE LEFT VENTRICLE

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Introduction: Limited data are available about global longitudinal strain (GLS) in patients with functional single ventricle, particularly single left ventricle (FSLV), and its clinical usefulness.
Purpose: To evaluate clinical utility of GLS compared to traditional echocardiographic parameters of systolic function in FSLV at pre Fontan stage and to identify potential correlation between GLS and haemodynamic data.
Methods: We retrospectively collected data about 22 consecutive patients from December 2011 to January 2016 (10 tricuspid atresia, 8 pulmonary atresia intact interventricular septum, 4 tricuspid and pulmonary atresia). Mean age at Glenn: 5 ± 50 months, mean age at pre Fontan catheterization: 54 ± 26 months. We identified two groups. Group 1 (13 patients): patients with haemodynamic findings adequate for Fontan completion who subsequently underwent successfully Fontan operation. Group 2 (19 patients): patients not eligible for Fontan completion or presenting early Fontan failure (2/19). Echocardiographic data analyzed were: LV ejection fraction (EF) by Simpson monoplane, four chamber 2D speckle GLS (4CGLS), mitral regurgitation. Catheterization data analysed included LV end-diastolic pressure and mean pulmonary pressure.
Results: There were no differences between groups in terms of LVEF ($59.7 \pm 4.5\%$ vs $57.5 \pm 3.7\%$, $p = 0.23$), LV end-diastolic volume (44 ± 27 ml vs 42 ± 20 ml, $p = 0.85$), mean pulmonary pressure (11.1 ± 1.2 vs 13 ± 2; $p = 0{,}7$), transpulmonary gradient (3.8 ± 1.2 vs 4.4 ± 1.6), mitral regurgitation. Significantly lower 4CGLS ($-25.7 \pm 3.1\%$ vs $-20.8 \pm 5.1\%$; $p = 0.01$) was found in Group 2 associated with higher LV end-diastolic pressure (7.1 ± 1.2 vs 9.1 ± 1.9; $p = 0.01$). Patients with worse 4CGLS showed a mild tendency to have higher LV end diastolic pressure ($R = 0.29$; $p = 0.19$).
Conclusion: Despite comparable values of LVEF and mitral regurgitation, patients not eligible for Fontan or with early Fontan failure had significantly lower 4CGLS compared to subjects who successfully underwent Fontan completion. 4CGLS might play a role in FSLV assessment and provide useful non-invasive information in pre Fontan assessment.

P1494 - CORONARY IMAGING UTILIZING GADOLINIUM CARDIAC MAGNETIC RESONANCE COMBINED WITH AN INVERSION RECOVERY GRADIENT ECHO SEQUENCE UTILITY IN CONGENITAL HEART DISEASE

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Background/Hypothesis: Coronary imaging in congenital heart disease and particularly in pediatrics can be challenging. We hypothesized that utilizing cardiac magnetic resonance (CMR) with an inversion recovery gradient echo sequence (IR-GRE) combined with gadolinium would be successful in visualizing the coronary arteries in the vast majority of cases.
Materials/Methods: All patients who presented for CMR and underwent IR-GRE between October 2010 and September 2016 were included. Gadolinium was either "dripped" in over the first third of the sequence or, if utilizing gadofosveset, administered as a bolus and IR-GRE performed immediately afterwards. Indications were either for coronary imaging or for imaging other structures (eg aorta) and coronaries were visualized.
Results: 1044 patients were found (54% male, age 14.3 + 12.6 years, range 3 days - 44 years) with 720 < 19 years. A whole range of diagnoses were included such as hypoplastic left heart syndrome, tricuspid atresia, transposition of the great arteries, bicuspid aortic valve, suspicion of anomalous coronary to name a few. Proximal coronary arteries were visualized in 987 (94.6%) – see figure. IR-GRE was successful even in patients with braces artifact where routine coronary imaging would fail. Myocardial scarring was able to visualized as well in the same exam – see figure. Of the remaining 57 patients, 28 (49%) were <1 year old and 13 (23%) were teenage males; success rate in the pediatric age range was 94%. 117 (12%) presented with a suspicion of an anomalous coronary of which anomalous right coronary from the left sinus was the most common finding ($N = 85$, 72.6%).
Conclusions: CMR utilizing IR-GRE combined with gadolinium has a high success rate (94.6%) in visualizing the coronary arteries in congenital heart disease and in the pediatric age range (94%); myocardial scarring was also visualized. Most failures were due to age <1 year or being a teenage male.

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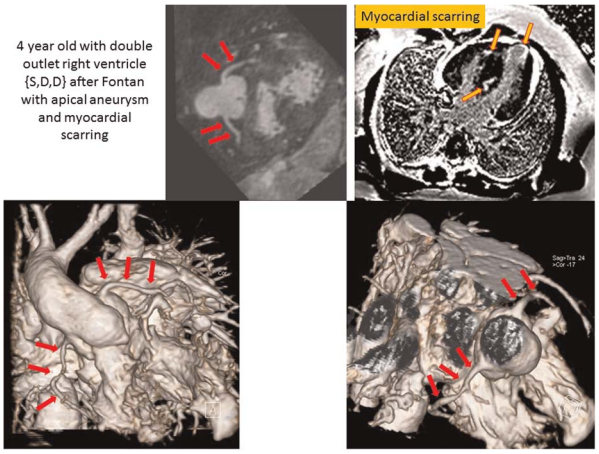

P1497 - SINGLE CORONARY ARTERY ARISING FROM PULMONARY ARTERY IN A PATIENT WITH DOUBLE OUTLET RIGHT VENTRICLE

Belen Toral[1], Ana Caro[2], Marta Flores[2], Miguel Angel Granados[2], Leticia Albert[2], Constanza Liebana[3], Miguel Rasero[3], Lorenzo Boni[4], Marta Olmedilla[5], Laura Diaz[5]
Hosptal Universitario 12 Octubre, Pediatric Cardiology, Madrid-Spain[1]; Hospital Universiatrio 12 Octubre, Pediatric Cardiology, Madrid-Spain[2]; Hospital Universiatrio 12 Octubre, Radiology, Madrid-Spain[3]; Hospital Universiatrio 12 Octubre, Cardiac Surgeon, Madrid-Spain[4]; Hospital Universiatrio 12 Octubre, Picu, Madrid-Spain[5]

Background: Anomalous origin of a single coronary artery form the pulmonary artery (ASCAPA) is a rare anomaly, described in literature as isolated clinical cases. We report a patient with postnatal diagnosis of ASCAPA, based on echocardiographic and CT images.
Materials and Methods: A female fetus was referred at 20 weeks' gestation with diagnosis of tetralogy of Fallot. At 40 weeks' gestation caesarean section was planned. Postnatal echocardiography showed double outlet right ventricle (DORV) with mild pulmonary stenosis, and biventricular dysfunction. Great arteries had a side-by-side arrangement. As for coronary arteries anatomy, we could only detect one coronary artery arising from the posterior wall of the main pulmonary artery, with an interarterial course. No right coronary artery was seen emerging from the aortic root. To complete the diagnosis a cardiac CT scan was performed using a 64 slice CT scanner. A retrospective, gated cardiac CT angiography was performed at 125 bpm. Analysis of the coronary arteries showed a single coronary artery (SCA) arising from de left and posterior side of the pulmonary trunk. The SCA bifurcated after a short course into a right coronary artery and a left main coronary artery. With these findings, surgery for translocation of the SCA was planned. The preoperative findings were confirmed intraoperatively and reimplantation of the SCA into the aorta was successfully performed using the double-flap technique. Postoperative course was complicated by right ventricular dysfunction and severe aortic regurgitation, requiring valvular replacement. DORV was surgically addressed 3 months later.
Conclusions: We present a rare case of ASCAPA associated to DORV. Fine diagnosis of coronary system is fundamental for a successful surgical treatment. Echocardiography alone could not rule out a dual coronary artery system, which was finally possible thanks to the actual advances in cardiac CT imaging techniques.

P1523 - ANALYSIS OF THE COMPOSITION OF CORONARY ARTERIAL CALCIFICATION POST KAWASAKI DISEASE WITH SINGLE SOURCE DUAL ENERGY CT

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In recent years, compositions of materials can become to be analyzed with dual-energy computed tomography (DECT), the effective atomic number (EAN) of coronary arterial calcification (CAC) was close to one of calcium oxalate monohydrate in adults. In this study, we examine the compositions of calcification of coronary arterial lesions of Kawasaki disease (KD-CAL) using EAN.
Object and Method: We had undergone DECT in 11 patients with KD-CAL. Exclusion criteria included patients who did not have a calcification more than 1mm and ones whose raw data had been erased. Age at KD was 0-6 years old (median 3 years old) and one at imaging was 15-45 years old (median 36 years old). DECT was performed with a prospective ECG-gated step and shoot technique. We reconstructed the field of view to 6.0 cm of the images and placed regions of interest (ROI) of 0.7mm diameter within the calcification 3 per 1 CAL at random. We calculated the distributions of the mean, median and maximum EANs, and compared the EANs with clinical records in every ROI.
Result: We obtained 69 EANs from 23 CALs. The median was 13.27 ± 0.83, the mean was 13.24 ± 0.80, the maximum was 13.59 ± 0.72, and these were close to the EAN of CAC. The analysis of every ROI, there were significant differences between age of KD (<1, 13.77 ± 0.52 vs $\geqq 1$, 13.16 ± 0.80, $p < 0.01$), time from onset of KD (<32, 13.69 ± 0.59 vs $\geqq 32$, 13.15 ± 0.80, $p < 0.01$) and BMI (<22, 13.70 ± 0.47 vs $\geqq 22$, 13.06 ± 0.88, $p < 0.01$).
Conclusion: The compositions of calcification of KD-CAL were close to ones of CAC. It was suggested that compositions of calcification of KD-CAL were different in age of KD, time from onset of KD and BMI. It is necessary to be careful about the onset of CV events.

P1531 - THE STUDY OF NEONATES WITH ISOLATED ABERRANT RIGHT SUBCLAVIAN ARTERY

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Background: The left aortic arch with an aberrant right subclavian artery (aRSCA) is rare but the most common congenital anomaly of the aortic arch. Although most cases of isolated aRSCA are asymptomatic, there are few reports concerning the clinical manifestations of neonates with isolated aRSCA.
Methods: A total of 1737 fullterm neonates, 900 males and 837 females, were received echocardiographic screening between January 2014 to March 2016. Among these patients, neonates with or without isolated aRSCA were studied, including the demographic data, perinatal history and clinical manifestations were recorded and analyzed during infant period.
Results: A total of 15 (0.9%) female predominant neonates, 6 males and 9 females, had isolated aRSCA (group 1) who were compared with 20 normal neonates (group 2). There were no significant difference in maternal age, gestational age, para gravity and gender in both groups but smaller baby's size (birth length and weight, $p = 0.023$ and 0.025, respectively) in Group 1. Although there

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were no significant difference in clinical manifestations, including developmental history, gastrointestinal and respiratory symptoms, neonates with aRSCA had higher incidences of mild developmental delay and feeding difficult than normal neonates (21% vs 0%, p = 0.061; 36% vs 20%, p = 0.264, respectively). The growth rates of body length and weight for age during the infant period were not significantly different between both groups.
Conclusions: The incidence of neonates with isolated aRSCA was low. They had smaller size at birth and some had mild developemental delay and feeding difficult, but no significant different growth rates.

P1548 - THREE-DIMENSIONAL ECHOCARDIOGRAPHY DATA IN THE AV VALVE SURGICAL ANATOMY CONSIDERATIONS IN PATIENTS WITH FUNCTIONAL SINGLE VENTRICLE

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Background: The atrioventricular valves regurgitation is one of the most significant risk factors worsening surgery outcomes in patients with univentricular hemodynamics. Three-dimensional echocardiography, applied since 2008 in our series, makes the anatomy of the atrioventricular valve and further intervention more clear. The goal of the study is to determine the possibility and benefits of the presurgery three-dimensional echocardiography in patients with functional single ventricle and atrioventricular valve regurgitation.
Materials and methods: An advanced transthoracic three-dimensional echocardiography systemic atrioventricular valve reconstruction protocol was applied in 43 patients with functional single ventricle of different types before or after surgery. The age of patients ranged from 3.2 to 30 yrs. The anatomy and function of the leaflets, the ring size, the degree and location of atrioventricular valve regurgitation was assessed in three-dimensional mode and compared with intraoperative data. 28 patients underwent atrioventricular valve regurgitation correction: 7 at the stage of the BCPC, 20 - at the Fontan procedure, 1 - after the Fontan procedure.
Results: All patients survived early and late after surgery. Three-dimensional echocardiography reconstruction data matched intraoperative anatomy of the atrioventricular valve in 96.5% of cases and supported preoperatively planned effective surgery. In 1 case the atrioventricular valve replacement was needed instead of the planned reconstructive surgery.
Conclusion: Three-dimensional echocardiography method allows you to get a clearer idea about the anatomy of the atrioventricular valve, his work, to determine the localization of regurgitation zones. The data obtained in most cases allow to plan upcoming volume method and the atrioventricular intervention valve. Three-dimensional reconstruction of the AV valves in patients with functional single ventricle can be recommended for inclusion in the standard echocardiographic examination protocol.

P1579 - THE EVALUATION OF PATENT DUCTUS ARTERIOSUS MORPHOLOGY IN DUCT DEPENDENT PULMONARY CIRCULATION USING THREE DIMENSIONAL COLOR FLOW ECHOCARDIOGRAPHY "ECHOCARDIOGRAM ANGIOGRAM"

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Background: Computed Tomogram angiography (CTA) is usually performed to delineate patent ductus arteriosus (PDA) tortuosity morphology in duct-dependent pulmonary circulation (DDPC) if two-dimensional echocardiography fails to provide PDA morphology information. Recently three-dimensional echocardiography color flow (3DECF) is able to create three dimensional color object "echocardiogram angiogram" to visualize a large degree scale in a single image.
Objective: To evaluate the relationship and accuracy of 3DE color flow in the assessment of PDA size and tortuosity in comparison to CTA. Methodology: A prospective study was performed from November 2014 until November 2015. Random selection involved 26 patients with DDPC who underwent CTA for PDA assessment. All patients underwent 3DECF study within 24 hours of CTA procedure. PDA was assessed for tortuosity and size of ampulla and the pulmonary end. The 3DECF data was compared against CTA which was blinded to the operator.
Results: Offline analysis of measurement PDA size was feasible in all patients, however delineating tortuosity of PDA with suppression tissue could be determined in 25/26 patients (96%). There was no significant difference between 3DECF and CTA in measurement of size pulmonary end (0.51 mm ± 0.14; 0.52mm ± 0.11, P-value = 0.92) and ampulla PDA (0.29mm ± 0.08; 0.27mm ± 0.97, P-value = 0.5) .There was a good correlation between 3DECF and CTA in determining tortuosity of PDA (r = 0.945) with inter-rater agreement K = 0.649. The 3DECF was highly accurate in detecting curved PDA (Sensitivity 100%; Specificity 100%) than detecting tortuosity with more than two curves (Sensitivity 88%; Specificity 100%).
Conclusion: 3 DECF is comparable to CTA in delineating the PDA size and tortuosity. However, the development of microarray transducer 3DE need to be improve for less stitching effect in acquisition full volume.

P1587 - TRENDS IN THE NUMBER OF NEW CASES OF ENDOMYOCARDIAL FIBROSIS SEEN IN THE UGANDA HEART INSTITUTE A TEN YEAR REVIEW

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Background/Hypothesis: Endomyocardial fibrosis (EMF), a neglected tropical disease, has historically been an endemic disease in Uganda. Recent reports from Nigeria and India have reported a reduction in new cases, leading to the theory that EMF is a vanishing disease. There is no data on epidemiological trends of EMF in Uganda. We aimed to fill this gap, by describing the trend of new pediatric EMF cases diagnosed at the Uganda Heart Institute (UHI).
Materials and Methods: We conducted a retrospective chart review of echocardiographic reports for all patients (aged <18 years) diagnosed with EMF who were seen in the division of Paediatric Cardiology of the UHI from January 2007 to December 2016. Patients presenting for repeat follow up evaluation were excluded. All transthoracic echocardiograms were reviewed by Paediatric cardiologists.
Results: A total of 102 cases of pediatric EMF were seen at the UHI during the ten years. Males comprised 59.8% (61/102) of cases. The mean age of the study population was 11.7 years (SD 3.3 years), with the youngest patient being 16 months old. Right

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Ventricular EMF was the most common form seen in 67.6% (69/102) of patients, the remaining being cases of biventricular EMF (most of which had predominant right ventricular involvement). No case of pure LV involvement was seen. Intracardiac thrombi were seen in ten cases. Pulmonary hypertension was documented in only 4 cases (3.9%), all of whom had biventricular EMF with significant mitral regurgitation. The total annual number of new cases of EMF declined over the period (n = 17 in 2007; n = 6 in 2016, see table 1).
Conclusions: There was a progressive decline in the number of new cases of EMF diagnosed at the UHI each year during the study period. This trend is similar to that observed in some parts of the world.

Table 1. Number of New Pediatric EMF Cases by Year over the Study Period

Year	2007	2008	2009	2010	2011	2012	2013	2014	2015	2016
EMF Cases	17	20	13	9	10	15	5	3	5	6

P1588 - CARDIAC CATHETERISATION UNDER MRI GUIDANCE USING A PARTIAL SATURATION APPROACH IN PATIENTS WITH CONGENITAL HEART DISEASE

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King, Division of Imaging Sciences & Biomedical Engineering, London-United Kingdom[1]; Evelina London Children, Paediatric Cardiology, London-United Kingdom[2]; King, Medical Physics, London-United Kingdom[3]; King, Medical Physics and Metrological Information Technology, Berlin-Germany[4]

Background: MRI represents a promising alternative to fluoroscopy to guide cardiac catheterisation. However, simultaneous high contrast visualization of catheter, soft tissue and blood remains challenging. In this study, we evaluated a novel passive tracking sequence based on partial pre-saturation (pSAT) for MR-guided catheterization in patients with congenital heart disease.
Methods: The technique was initially tested using a 3D-printed phantom. Two patients (12 and 39 years-old) in whom an XMR was indicated for pulmonary vascular resistance assessment were recruited. Underlying diagnosis were severe right pulmonary stenosis and atrioventricular septal defect (child and adult, respectively). MR-guided catheterization was performed using a real-time single shot acquisition with bSSFP readout (TR/TE = 2.6 ms/1.3ms,flip angle = 60°, FOV = 370 × 370mm2,voxel size = 2.2 × 2.5mm2,bandwidth = 1190 Hz,SENSE factor = 2.5, partial Fourier = 0.65,acquisition time = 145ms,linear ordering). The balloon of the wedge catheter was filled with 1%Dotarem® for positive contrast visualization. A partial saturation prepulse (modified flip angle to 30°) was applyed before the acquisition of MR-images. The sequence was run in interactive mode, the imaging plane location was modified in real time using pre-programmed pedals.
Results: The balloon of the wedge catheter was clearly depicted during navigation in the phantom with the pSAT sequence(Figure 1). Right heart catheterization was performed in both patients (Figure 2). The pSAT sequence was successful to passively track the catheter and simultaneously visualize soft tissue and blood. However, in the child, the catheter lacked the stiffness required to cross the pulmonary valve, therefore the procedure was completed with X-ray support using a braided non-MRI compatible catheter over a wire. The procedure was completed solely under MRI-guidance in the adult patient. Total procedure time was 172 and 170 minutes, cardiac catheterisation time was 55 and 25 min in child and adult, respectively.
Conclusions: The proposed pSAT sequence provides simultaneous high contrast visualization of the catheter balloon, soft tissues and blood. This technique provides excellent passive tracking capabilities during MR-guided catheterization.

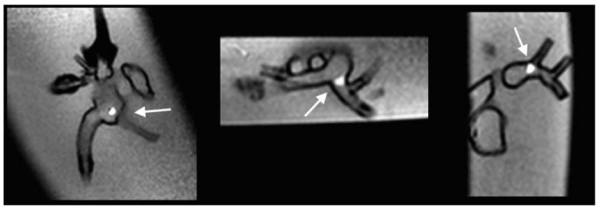

Figure 1.
Example real-time images acquired in the 3D printed heart phantom with balloon wedge catheter using the pSAT sequence. The catheter tip (arrows) could be easily tracked and visualized in different imaging plane orientations.

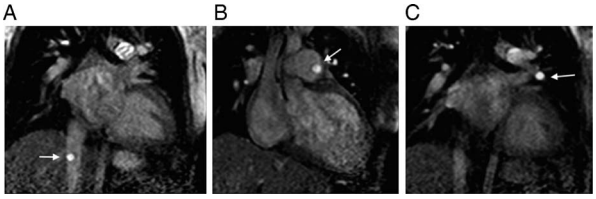

Figure 2.
Real-time images acquired in the adult patient using the pSAT sequence. Note balloon of the wedge catheter (arrows) at the inferior vena cava (A), main pulmonary artery (B) and left pulmonary wedge (C). The pSAT sequence provided excellent and simultaneous visualization of catheter tip, blood and soft tissues.

P1598 - SENSITIVITY AND SPECIFICITY OF FETAL ECHOCARDIOGRAPHY IN HIGH RISK PREGNANCIES A SINGLE CENTER EXPERIENCE FROM EGYPT

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Background: Advances in fetal Echocardiography training among pediatric cardiologists has led to improved antenatal detection of congenital heart diseases. Nevertheless, this subspecialty is still evolving in developing countries. No research has determined fetal echocardiography sensitivity and specificity in developing countries like Egypt.
Subject and Methods: Study was a retrospective analysis of pregnant females referred to cardiology unit in Mansoura university children hospital, Egypt, for fetal echocardiography according to indications of American Heart Association. All cases who fulfilled the indications and had detailed fetal and neonatal echocardiogram were included. Results of fetal and postnatal echocardiography were compared to assess sensitivity, specificity of fetal Echocardiography. Minor lesion was defined when no postnatal

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intervention is needed and Major when postnatal intervention was scheduled.
Results: Out of 227 referred cases from January 2011 to January 2016, comparisons were possible in 164 fetuses as 63 were either dead (stillborn or terminated) or lost follow up. Mean age of ladies included in the study was 26.64 ± 5.428 years and mean gestational age at referral was 27.59 ±5.41 weeks ranging (16-39 weeks). Sensitivity and specificity calculation for fetal echocardiography were performed by comparing fetal and neonatal diagnoses. Cases had minor differences (n = 3), 2 fetuses were diagnosed as VSD but normal postnatal and one normal prenatal was proved to have mild pulmonary stenosis postnatal. Prenatal diagnosis in our study was accurate in 98.17% of cases but accuracy in diagnosing major lesions 100%. Sensitivity of fetal echocardiography in our study was 98.64% and specificity of 97.78%.
Conclusion: Fetal echocardiography is considered a sensitive and specific tool for prenatal congenital heart diseases detection even in developing country like Egypt. Obstetricians' awareness in developing countries should be raised regarding the high accuracy of fetal echocardiography in high risk pregnancies to guide antenatal and early neonatal management of congenital heart diseases.

P1609 - THE ARCHITECTURE OF THE RIGHT VENTRICULAR MYOCARDIUM CHANGES THROUGH THE CARDIAC CYCLE

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Background: Right ventricular heart failure constitutes a significant clinical challenge in several patients with congenital heart disease. Changes in the myocardial structure has recently been suggested as a contributing factor to heart failure. However, no studies have investigated the normal three-dimensional rearrangement of the cardiomyocytes through the cardiac cycle in the healthy right ventricle. In the present study, we have investigated these dynamic changes by the use of diffusion tensor magnetic resonance imaging.
Methods: 14 healthy female domestic 20 kg pigs were included. The hearts were excised and randomized to fixation in either diastole or systole. The orientation of the cardiomyocytes was quantified using diffusion tensor magnetic resonance imaging and compared between systole and diastole.
Results: From diastole to systole the orientations of the cardiomyocytes changed most pronounced in the endocardium. In both ventricles, the cardiomyocytes changed from a more circumferential orientation in diastole to a more longitudinal orientation in systole. In the right ventricle, the increase in angulation from diastole to systole was 6.45° [4.4; 8.6]° whereas this angulation change in the left ventricle was 8.2° [5.8; 10.6]°. The two ventricles differed with regards to change in transmural orientation of the cardiomyocytes. In the left ventricle, the transmural angulation increased from diastole to systole with 5.0° [4.2; 5.9]°. Cardiomyocytes located in the right ventricle showed no significant change in transmural orientation through the cardiac cycle.
Conclusion: The architecture of the entire myocardium changes through the cardiac cycle. This rearrangement is provided by continuous changes in the orientation of cardiomyocytes. The movement of cardiomyocytes is, however, different in the right and left ventricle indicating that the normal physiological remodelling of the myocardium differs between the ventricles. These results must be taken into account when assessing remodelling in the diseased myocardium, particularly in patients with right ventricular heart failure.

P1613 - ASSESSMENT OF EARLY ATHEROSCLEROSIS IN CHILDREN WITH WHITE COAT AND ESSENTIAL HYPERTENSION

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Introduction: Sonographic intima media thickness measurement of the carotic artery (cIMT) is considered a valid surrogate marker for cardiovascular risk allowing assessment of atherosclerotic changes at very early stage. Elevated blood pressure in adults is associated with increased cIMT. However, its effect on arterial health is less understood in a paediatric population. Thus, the aim of this study was to evaluate cIMT in adolescents suffering from essential and white coat hypertension.
Methods: One hundred thirty eight children – 46 (23 boys) children suffering from WCH, 46 (23 boys) patients with essential hypertension and 46 age/gender-matched healthy controls (age-period from 14 to 18 years) - were examined under standard conditions. The mean IMT of the carotid arteries was measured by high-resolution B-mode ultrasound (Prosound F75 Aloka).
Results: Statistical analysis revealed significant differences in the mean cIMT between the both hypertensive groups (WCH, EH) and control group (0,45 ± 0,05 mm for WCH; 0,47 ± 0,06 mm for EH vs 0,40 ± 0,04 mm; $p < 0.001$). No significant differences were found between WCH and essential hypertensive groups.
Conclusions: Our results revealed significantly higher mean cIMT in children suffering from white coat and essential hypertension indicating early atherosclerotic changes associated with increased cardiovascular risk already in adolescent hypertension. Importantly, WCH should not be considered a harmless trait and has common features with essential hypertension.
Support: VEGA 1/0087/14, project "Biomedical Center Martin" ITMS code [26220220187] co-financed from EU sources.

P1635 - MITRAL VALVE PERFORATION AMONG CHILDREN SEEN AT THE UGANDA HEART INSTITUTE A SERIES OF NINE CASES

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Background: Mitral valve perforation is a rare complication of infective endocarditis (IE), especially in those receiving inadequate therapy. In Africa rheumatic valvular heart disease (RHD) is a major underlying cause of IE. The perforation of mitral valve worsens any pre-existing mitral regurgitation (MR) and hence is associated with increased mortality. There are few reports on mitral valve perforation from sub-Saharan Africa.
Objectives: We describe the clinical presentation and outcomes of children with echocardiographically diagnosed mitral valve

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perforation followed up at the Uganda Heart Institute (UHI) from July 2011 to June 2013.
Methods: We reviewed the clinical and echocardiographic data of patients with mitral valve perforation, defined as MR with multiple jets, an area of echolucency on anterior mitral leaflet (AML) or perforated aneurysm on the AML.
Results: A total of nine patients were diagnosed to have mitral valve perforation, 8 being female. The median age was 12 years (range 15mo-14years). None had prior cardiac surgery, penetrating chest trauma, cardiac catheterization or radiofrequency ablation. All patients were treated for a long standing febrile illness suggestive of IE prior to cardiology referral. Blood cultures were positive in only one patient which grew Streptococcus viridans. Two patients had no predisposing heart lesion on echo, while the rest had underlying RHD. All patients had severe MR through multiple jets while one patient had additional severe aortic regurgitation. Two patients had visible vegetations on the AML. Two patients had a ruptured AML aneurysm. Four patients (44.4%) died within two months of diagnosis. None had heart surgery since this was not routinely available at UHI at the time.
Conclusion: RHD is a major underlying cause of IE and subsequent mitral valve perforation in our settings. Many patients with mitral valve perforation have high immediate mortality.

P1637 - AN ANALYSIS OF VOLUME TIME CURVES (VTC) OF THE LEFT VENTRICLE (LV) BASED ON FAST FOURIER TRANSFORMS (FFT)

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Objectives: For the measurement of LV volume, 3D real-time Echocardiography (3DRTE) has been established as an accurate and safe method. As a result, the volume can be computed and plotted as a time-dependent signal, which is defined as a VTC. The VTC allows the analysis of the systolic and diastolic function. However, these signals are difficult to analyze and process. Aim of our study is to define mathematical functions of the VTC, which allow a better quantification, assessment and comparison of the systolic and diastolic function.
Methods: In 320 healthy and 20 children with congenital heart disease, 3DRT datasets of LV were acquired using the iE33 ultrasound scanner (Philips; transducer X7-2 and X5-1). 3D datasets were segmented with ImageArena (TomTec), a semi-automatic software tool. We used Python for the following data processing. The resulting VTC was used to reconstruct the volume curve for multiple heart beats, assuming a steady state. The missing data were interpolated. We used the FFT to decompose the signal into a representative sum of trigonometric functions. For an analysis, we use the amplitude spectrum of the VTC, shown in figure 1.
Results: We successfully implemented the FFT and used it to compare the VTC of healthy and pathological hearts. In an experiment, we showed that a healthy VTC superimposed with noise can be detected in the diagram of the amplitude spectrum. The noise represents a malfunction of the heart and is modelled as sinus wave in our experiment. Consequently, the noise can be detected and methods can be adjusted to eliminate malfunctions.
Conclusions: The FFT is a crucial method to analyze VTC and to derive information how a cardiac dysfunction could be treated. The decomposition of VTC into mathematical function can be useful to customize medications, plan interventions or configure pacemakers.

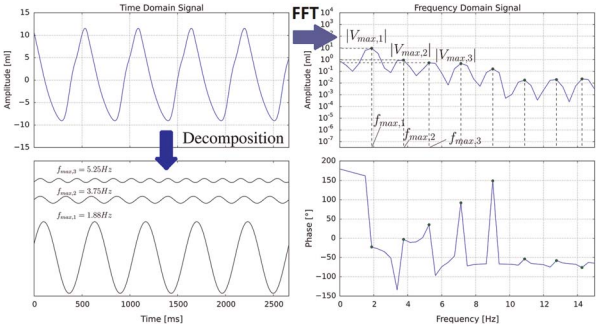

Figure.

P1644 - EVOLUTION OF BACTERIAL ENDOCARDITIS OF MITRAL VALVE IN CHILDREN UNDER ONE YEAR

Alberto Robredo
Children and Women Hospital, Cardiology, Salta-Argentina

Background: The significant insufficiency and prolapse of the mitral valve (MV) is a rare condition in infants. The early diagnosis of bacterial endocarditis (BE) in this condition requires a great clinical suspicion at this age. The objective is to show the unfavorable evolution of three cases of BE on MV with prolapse and insufficiency in children under one year.
Material and Methods: We analyzed BEs on MV in the last 5 years: 1) Male 7 months old with a history of 2 months before surgery of aorta and ductus coarctation with very good evolution. He entered with sepsis and heart failure in respirator. He was diagnosed with BE caused by wart in MV with severe insufficiency and positive blood cultures. He died due to multiorgan failure. 2) Female, 8 months old without history, enters with sepsis. Diagnosed by echocardiogram with Myxomatous MV with prolapse and severe insufficiency and a wart of more than 1 cm. Blood cultures negative. She presents cerebral embolism with severe encephalic compromise. The MV was replaced with unstable evolution and died 6 months later. The diagnosis of BE was through pathological tests of surgical samples. 3) Male, 2 months old, presents BE with positive blood cultures and wart in Myxomatous MV by echocardiogram. He presented poor evolution due to associated factors. None of them had a proved entrance.
Results: All three patients had Myxomatous MV with prolapse and severe insufficiency with unfavorable evolution of the BE despite the efforts made.
Conclusions: It is advisable a thorough exploration in infants to search insufficiency and prolapse of MV, due to the bad evolution of BE, for a further strict prophylaxis compliance.

P1671 - TRANSIENT DIASTOLIC DYSFUNCTION IN ACUTE PHASE OF KAWASAKI DISEASE WITHOUT CORONARY ARTERY LESION

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Niigata City General Hospital, Pediatrics, Niigata-Japan[1]; Okinawa Prefectural Nanbu Medical Center & Children's Medical Center, Pediatric Cardiology, Okinawa-Japan[2]

Impaired left ventricular (LV) systolic function during acute phase of Kawasaki disease (KD) without coronary artery lesion has been described in some reports. Our purpose of this study is to investigate LV diastolic function in acute phase of KD. We retrospectively studied 82 KD patients admitted to Niigata City

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General Hospital from August 2013 to December 2015. Mean age of the patients at admission was 2.8 + /- 2.1 years, and 68% were boys. All the patients underwent transthoracic echocardiography by just one echocardiographer in acute phase (AP) (10.8 + /- 2.0 days of illness) and in recovery phase (RP) (49.1 + /- 9.8 days of illness), and they were administered intravenous immunoglobulin before the first echocardiography. We compared echocardiographic measurements in AP and RP, which contained LV diastolic dimensions (LVDd) transformed to z-scores, LVEF by Pombo's equation, isovolemic relaxation/contraction time (IVRT/IVCT), total ejection isovolemic index (Tei index), the early mitral inflow velocity to interventricular septum annular early diastolic velocity (IVS E/e'), the early mitral inflow velocity to lateral mitral annular early diastolic velocity (LV E/e'), and the early tricuspid inflow velocity to tricuspid annular early diastolic velocity (RV E/e'). Differences were evaluated by t-tests and statistical significance was achieved with p value < 0.05. Ten of 82 patients had transient coronary artery dilatation, but all of the lesions were regressed. IVS E/e', LV E/e', Tei index, and IVRT were significantly increased in AP compared with RP ($p = 0.005$, $p < 0.001$, $p = 0.03$, and $p = 0.004$, respectively). There were no significant differeces in LVDd, LVEF, IVCT, and RV E/e'. In conclusion, echocardiographic parameters which indicate LV diastolic function were worse in about 10 days than 50 days of illness in KD. These paremeters of diastolic function may be more sensitive than those of systolic function in subtle myocardial injury in acute phase of KD.

P1678 - PERSISTENT PERICARDIAL EFFUSION IN A NEWBORN. A CLUE FOR EXRALOBAR PULMONARY SEQUESTRATION

Timur Meşe[1], Murat Muhtar Yilmazer[1], Rahmi Özdemir[2], Bariş Güven[3], Aysun Hacer Sritaş[4]
Izmir Dr.Behçet Uz Children's Hospital, Pediatric Cardiology, Izmir-Turkey[1]; Dumlupinar University Evliya Çelebi Training and Research Hospital, Pediatric Cardiology, Izmir-Turkey[2]; Tepecik Training and Research Hospital, Pediatric Cardiology, Izmir-Turkey[3]; Izmir Dr. Behçet Uz Children's Hospital, Pediatrics, Izmir-Turkey[4]

Introduction: Here we report an unusual case of an pulmonary sequestration diagnosed was made through persistent and localised pericardial effusion.
Case: Twenty five days-old boy reffered from neonatology outpatient clinic for the evaluation of a grade 1/6 systolic murmur. His physical cardiac examination and 12-lead-electrocardiogram was normal. Chest x-ray revealed cardiothoracic ratio with upper limit of normal and highligtened thymus. Echocardiographic examination revealed normal anatomic cardiac structure and function with an unusual local pericardial effusion on the lateral border of left ventricle (4 mm in systole and 3.6 mm in diastole). Although the child had no sign of respiratory or cardiovascular signs sent home to visit again. After a week pericardial effusion was persisting with no oblivious change in the effusion volume. His second chest x-ray was also reported by radiologist as normal. Cardiac MR was taken to visualize the detailed anatomy of cardiac structure. A MRI confirmed the slight pericardial effusion on lateral border of left ventricle. In addition consolidated inferior left lobar pulmonary area was noted and computed tomography (CT) evaluation was also suggested. A CT scan of the chest with intravenous contrast showed triangular shaped mass in the left lower lobe without pleural effusion (Figure 3. CT1). An aortic branch was directed to the extra lobar sequestrated pulmonary area (Figure 3. CT2).
Conclusions: Pulmonary sequestration has the incidence of %0.1. About %15-25 of extrapulmonary form constitude sequestration with main differential structure is having its own pleura and mostly situated on left lung. Most of the extrapulmonary sequetrations presented in neonatal period with infection or cyanosis. Localised persistent pericardial effusion must be evaluated rigoriously for the etiologial aspects especially in newborns and congenital pulmonary pathologies must be keep in mind.

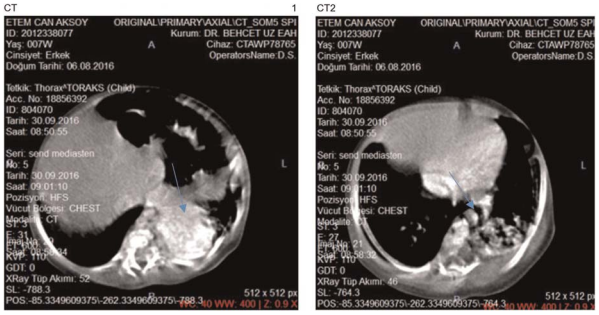

Figure.

P1683 - CARDIAC LIPOMA

Susana Uriel[1], Gloria Sánchez[2], Noemí Cañete[2], Dimpna Albert[1], Marc Figueras[3], Stella Pie[1], Lluís Mayol[1], Raúl Abella[4], Ferran Rosés[3]
H. Dr J. Trueta, Paediatrics, Girona-Spain[1]; H. Dr J.Trueta, Radiology, Girona-Spain[2]; H. Vall Hebrón, Paediatrics, Barcelona-Spain[3]; H. Vall Hebrón, Cardiac Surgery, Barcelona-Spain[4]

Background: Cardiac lipomas are uncommon benign primary cardiac neoplasms. Typical locations are the endocardium of the right atrium and the left ventricle. They are usually asymptomatic but symptoms, when present, depend on the location within the heart and the tumor's size. Diagnostic modality of choice is cardiac MRI. Treatment guidelines have not yet been established due to the very low prevalence of these tumors and are thus guided by the patient's symptomatology.
Materials and Methods: We present a case of a twelve year old boy with a non invasive and asymptomatic cardiac lipoma which was detected as an electrocardiogram anomaly. An echocardiogram performed which revealed an hyperechoic mass on the left side of interventricular septum (see Figure 1). Then a cardiac MRI was performed for better tissue characterization The MRI revealed a long, fat containing mass next to the interventricular septum, without extension or invasion into other cardiac parts. The mass appeared to represent a lipoma base on its imaging characteristics (see Figure 2).
Results: The case was discussed by a team of cardiologists and cardiacs surgeons, which recommended to the family wait and see due to the lack of in the patient symptoms, and the high biopsy risk based the tumor's location. No increase in tumor size was detected in the following cardiac MRI and no arrhythmias appeared on the follow up (one year). The patient is still asymptomatic.

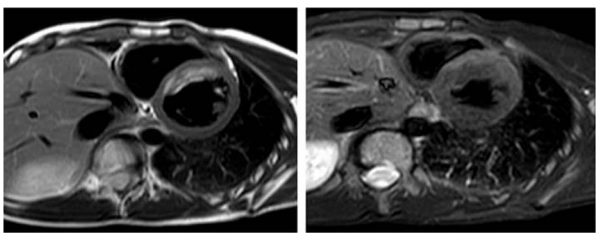

Figure 1.

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Conclusions: Cardiac lipomas are rare benign tumors of the heart which are usually asymptomatic. Diagnostic modality of choice is cardiac MRI. Treatment guidelines have not yet been established, but are guided by the patient's symptomatology. Follow up is recommendable.

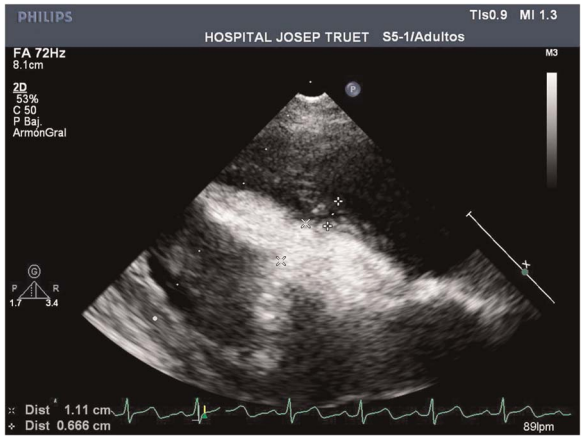

Figure 2.

P1687 - RIGHT CERVICAL AORTIC ARCH PRESENTING WITH A PULSATILE NECK MASS IN A CHILD

Murat Muhtar Yilmazer[1], Timur Meşe[1], Rahmi Özdemir[2], Bariş Güven[3]
Izmir Dr. Behçet Uz Children's Hospital, Pediatric Cardiology, Izmir-Turkey[1]; Dumlupinar University Evliya Çelebi Training and Research Hospital, Pediatric Cardiology, Kütahya-Turkey[2]; Tepecik Training and Research Hospital, Pediatric Cardiology, Izmir-Turkey[3]

Case: A 9-year-old Syrian girl was referred to our pediatric cardiology clinic for evaluation of a pulsatile neck mass. She has been examined for Turner Syndrome due to short stature but chromosomal analysis demonstrated a 46 chromosomes present, and the XX. Physical examination revealed a prominent pulsation above the right sternoclavicular area.A systolic murmur, grade 1/6, in the aortic position was heard. Transthoracic echocardiography (TTE) revealed normal ventricular morphology and functions and tricuspid aortic valve with no regurgitations or stenosis. However images of the aortic arch could not be obtained with the usual suprasternal angulation. We were able to demonstrate the aortic arch by angulation of the probe slightly to the right. However branching of the aortic arch could not be demonstrated clearly. As TTE was insufficient for determination of detailed configuration of the aortic arch, cardiac catheterization and angiography were performed. Aortography in left lateral position demonstrated a high-lying aortic arch extending cranially to the toracic aperture (figure 1A). Normally the aortic arch lies behind the lower part of the manubrium sterni and does not exceed to the sterno-clavicular joint. In this case, a portion of the ascending aorta and the aortic arch exceeded the sterno-clavicular angle and extended to the neck. An angulation which was not create any obstruction in the aortic lumen shown in the descending aorta. Although the branching pattern of the aorta was usual, all 3 branches were thinner than normal (figure 1B). Pressure gradient were recorded in 3 major branches and no stenosis was detected. Computed tomographic angiography revealed a right cervical aortic arch extending to the neck and turned downward on itself to become the descending aorta which was the cause of angulation in descending aorta detected by angiography (figure 1C and 1D).

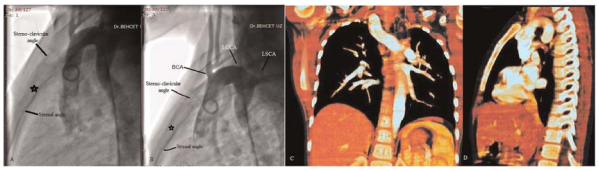

Figure.

P1690 - CHRONOLOGICAL SERIAL CHANGES OF SYSTOLIC AND DIASTOLIC FUNCTION BY USING TISSUE DOPPLER IMAGING (TDI) IN HEALTH NEONATES WITHIN THE FIRST 3 DAYS OF LIFE

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College of Medicine, Korea University, Pediatrics, Seoul-Korea, South

Background: Several significant hemodynamic changes by closure of PDA take place during transition from the fetal to neonatal environment. There are many echocardiographic researches assessed by TDI during transitional period but they have not represented serial changes during immediate postnatal period within 48 hours after birth. The goal of the present study was to evaluate delicate echocardiographic serial changes assessed by TDI parameters within 48 hours after birth in full term infants.
Materials and Methods: One-center, prospective, observational longitudinal cohort study in healthy newborns. They underwent standard echocardiography and TDI from 12 to 72 hours after birth and followed at 12 hours intervals during the first 3 days of life. Chronological cardiac morphometry, functional systolic and diastolic parameters including TDI were measured in LV and RV.
Results: 56 infants were measured serially two or three times with 12-hour interval. Changes of aortic VTI, closure of PDA, LV CO, MV and TV diastolic parameters, RV TDI E', A' and S', LV MPI and RV MPI, LV and RV EDV showed significant or insignificant differences between postnatal age before and after 24 h.
Conclusions: It is meaningful that our study departmentalized basic data of immediate changes of cardiac morphometries, functional, diastolic parameters, and TDI within postnatal 72 hours. Two parameters of postnatal age 24 h and PDA closure have an important effect on cardiac changes during immediate transitional period.

P1696 - PREDICTING THE NEED FOR SURGICAL REPAIR OF PRENATALLY DIAGNOSED COARCTATION OF THE AORTA

Conall Morgan[1], Brigitte Mueller[2], Callaghan Jull[1], Luc Mertens[1], Mark Friedberg[1], Vitor Guerra[1], Ra Han[1], Varsha Thakur[1], Mike Seed[1], Edgar Jaeggi[1], Cedric Manlhiot[2], Lynne Nield[1]
The Hospital for Sick Children, Labatt Family Heart Centre, Toronto-Canada[1]; University of Toronto, Cardiovascular Data Management Centre, Toronto-Canada[2]

Background: In prenatally diagnosed coarctation of the aorta, it is difficult to predict which neonates will require early surgical intervention.
Methods: From January 2005 to December 2015 fetal cases referred for suspected coarctation were reviewed. Patients with an apex forming left ventricle with antegrade flow across the mitral and aortic valves were included for analysis.

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Results: A total of 110 patients were identified. 3 patients (3%) suffered in-utero demise. Median gestational age at diagnosis was 31 weeks (IQR 23-35 weeks). Postnatally, 47 patients (44%) underwent a biventricular repair, 56 patients (52%) required no intervention and 4 patients (4%) underwent a staged biventricular repair. Area under the curve was 0.9013. In univariate analysis a hypoplastic transverse arch (OR 4.33 [1.92-10.3], p = 0.001), an increase in LVOT peak Doppler (OR 1.02 [1-10.3], p = 0.03), and an increase in pulmonary valve z-scores (OR 1.61 [1.18-2.32], p = 0.005) increased the odds of requiring postnatal surgical repair significantly. An increased isthmus diameter (OR 0.23 [0.11-0.42]/mm, p = < 0.001), arterial duct diameter (OR 0.64 [0.48-0.84]/mm, p = 0.002) and increased aortic (OR 0.72 [0.54-0.94/z, p = 0.02), mitral (OR 0.58 [0.42-0.78]/z, p = 0.001) and ascending aorta (OR 0.52 [0.38-0.7]/z, p < 0.001) z-scores significantly decrease the odds of requiring surgery postnatally in the univariate analysis. In multivariate analysis, an increase in LVOT peak Doppler (OR 1.05 [1.03-1.09]/cm per sec, p = < 0.001) increases the odds of requiring a surgical repair, while an increase in ascending aorta z-score (OR 0.60 [0.40-0.84]/z, p = 0.005) significantly decreases the odds of requiring a surgical repair postnatally.
Conclusions: Peak LVOT Doppler and ascending aorta z-score can help in predicting which fetuses will require early surgical intervention.

P1704 - MYOCARDIAL FISTULISATION AND CORONARY ARTERIAL ECTASIA IN CHILDREN WITH UNIVENTRICULAR CIRCULATION – AN UNDER RECOGNISED PROBLEM

Daragh Finn[1], Kevin Walsh[1], David Roberson[2], Colin Mcmahon[1]
OLCHC, Crumlin, Cardiology, Dublin-Ireland[1]; Advocate Christ Hospital and Medical Center, Oak Lawn, Il 60453, Cardiology, Naperville-United States[2]

We describe three children, each with a univentricular circulation, who had evidence of diffuse fistulisation of the myocardium at cardiac catheterisation. One of the children had documented normal coronary arteries and myocardium at catheterisation prior to development of the fistulisation process. The same child also developed extensive veno-venous collaterals. All children had evidence of elevated pulmonary arterial pressures and were treated with a combination of sildenafil and bosentan pulmonary vasodilators. The presence of chronic hypoxaemia, elevated filling pressures and the use of pulmonary aterial vasodilators could potentially have contributed to coronary endothelial dysfunction, thereby giving rise to the coronary vasculopathy. This may necessitate early referral for orthotopic cardiac transplantation. Diffuse fistulisation of the myocardium in children and young adults with univentricular circulation may be an under-recognised occurrence and coronary angiography should be considered in these patients.

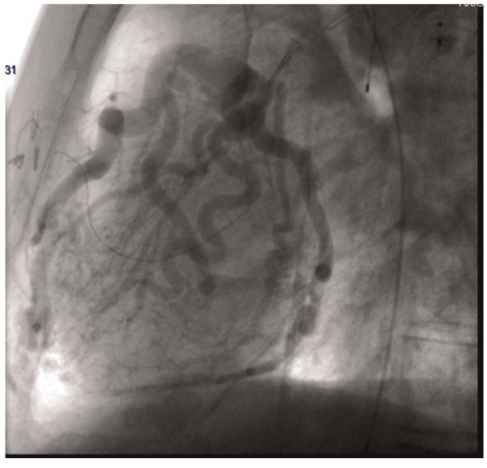

Figure.

P1715 - CARDIAC MAGNETIC RESONANCE AS A FOLLOW UP TOOL IN PATIENTS WITH GIANT CORONARY ARTERY ANEURYSMS SECONDARY TO KAWASAKI DISEASE IN A MEXICAN POPULATIO

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Background: Kawasaki's disease (KD) is an infancy systemic vasculitis that without treatment is associated with coronary abnormalities and cardiovascular damage in nearly 25% of the cases. Prognosis is directly related to the presence and severity of the coronary lesions. Cardiac Magnetic Resonance (CMRI) is a non-invasive tool used for an accurate determination of cardiac function in several pathologies.
Objective: To describe the findings in Cardiac MRI as an anatomic and functional evaluation in the long-term follow-up of patients with a history of KD and giant coronary aneurysms.
Material and Methods: Cross-sectional, observational and descriptive study of patients with history of KD and giant aneurysms (z-score >10) treated in the Instituto Nacional de Pediatria in Mexico City, Mexico from August 1995 to June 2015.
Results: We performed a CMRI in 23 patients with a mean age of 104.7 months (range 24 to 192 months). Mean time between diagnosis of KD and the cardiac MRI was 83.16 months. 16 patients were male (70%). We found abnormalities in the CMRI in 18 patients, with presence of coronary artery aneurysms in 12 cases. The ventricular function was abnormal in 12 cases, including 2 patients without coronary artery lesions.
Conclusions: CMRI is a useful tool in the follow-up in patients with KD showing not only the presence of coronary abnormalities, but also provides functional evaluation that could be present in patients without coronary aneurysms.

P1722 - CARDIAC MAGNETIC RESONANCE IN LONG TERM FOLLOW UP FOR TETRALOGY OF FALLOT CHILEAN EXPERIENCE

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Background: Tetralogy of Fallot (ToF) is the most common cyanotic congenital heart disease. Surgically repaired patients may present long-term complications: pulmonary regurgitation (PR) and right ventricle (RV) enlargement and dysfunction, amongst others. Cardiac magnetic resonance (CMR) is the gold standard for RV evaluation, but it is not widely available in our country.

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Objective: Describe CMR results in long-term follow-up of our ToF population that has no undergone pulmonary valve replacement (PVR).
Materials and Methods: All CMR performed at our institution between 2007 and 2012 in repaired ToF patients with no PVR were included. Pulmonary valve regurgitant fraction (RF), ventricular end diastolic (EDV) and systolic volumes (ESV), and ejection fraction (EF) were examined.
Results: 122 CMR were performed in 114 patients (59% male). Mean age at CMR was 15.4 ± 7.4 yo. Median age at surgery was 12.2 months-old (2-27 months-old). 78% of patients received transannular patch (TAP) and 22% underwent pulmonary valve-sparing repair. Severe PR (RF > 40%) was present in 53.3%. RVEDV was 157.3 ± 38.6 ml/m2 and RVESV was 85.3 ± 27 ml/m2 (median ± SD). RVEDV was >150 ml/m2 in 48.4% and >170 ml/m2 in 32.8% of patients. Patients with TAP showed larger RV volumes compared with those without TAP: RVEDV 165.6 ± 39.5 vs 133.6 ± 30.7 ml/m2 ($p < 0.01$) and RVESV 90 ± 27.7 vs 73.3 ± 24.8 ml/m2 ($p < 0.02$). Mean RVEF was 46.4 ± 7.1% and left ventricle EF 55.8 ± 7.6%. Systolic dysfunction was seen in 43.7% of patients for RV and 35.2% for LV. There was no difference in RVEF nor LVEF regarding surgical technique. Patients with RVEDV >170 ml/m2 showed worse RVEF that those with smaller RVEDV (47.9 ± 7% vs 43.2 ± 6.4%, $p < 0.01$).
Conclusions: Almost half of patients showed significant RV enlargement, demonstrating that our population is having CMR late in their follow-up. TAP was associated with larger RVEDV and RVESV, but no worse RVEF in this cohort. Larger RVEDV was associated with worse RVEF.

P1750 - ECHOCARDIOGRAPHIC FINDINGS IN CHILDREN WITH CONGENITAL ZIKA VIRUS

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Background/Hypothesis: The Zika virus (ZIKV) is an arbovirus, transmitted mainly by mosquitoes of the genus Aedes. An increase in children born with microcephaly was noted in 2015 in regions of Brazil with high transmission of ZIKV, particularly the northeast. Several malformations were reported in babies born with congenital ZIKV in the country since. The need to verify the virulence of Zika as the pivot of cardiac changes in children born with microcephaly was noted.
Methods: In this study, 40 infants clinically diagnosed with ZIKV-related microcephaly (cephalic perimeter <32 cm), were submitted to echocardiography (ECHO). Those with microcephaly of other known cause or presented debilitation culminating in failure to perform the ECHO were excluded.
Results: From the children with presumed or confirmed congenital ZIKV examined, there were 22 females. The mean age was 6.79 month (range 3 to 10 months). The cephalic perimeter at birth range from 25 to 32 cm - mean 28.5 cm. The gestational age of maternal ZIKV infection in 21 patients was the first trimester of pregnancy, 10 in the second and 4 in the third, 4 mothers were unable to report the period of infection. Thirty-six mothers referred symptoms during pregnancy such as fever (18), exanthema (36), arthralgia (20), myalgia (17), headache (19), cough (4) and conjunctival hyperemia (12). Regarding the ECHO exam, 12 patients had patent foramen ovale (PFO), 3 had small ostium secundum interatrial communication (ASD), one small patent ductus, one ventricular muscular septal defect (VSD) that closed spontaneously and 23 had normal ECHO.
Conclusions: Among children evaluated in this data, there was no association between ZIKV and complex congenital heart diseases. The echocardiographic findings were small defects without hemodynamic repercussions.

P1776 - SUBCLINICAL LEFT VENTRICULAR DYSFUNCTION IN JUVENILE DERMATOMYOSITIS PATIENTS A TWO DIMENSIONAL SPECKLE TRACKING ECHOCARDIOGRAPHIC STUDY

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Background: Little is known about left ventricular (LV) function in pediatric patients with juvenile dermatomyositis (JDM), especially in asymptomatic individuals with preserved ejection fraction (EF). The study aimed to identify subclinical LV systolic and diastolic dysfunction in JDM using two-dimensional speckle-tracking echocardiography (2DST).
Methods: 20 consecutive JDM patients without cardiac symptoms were enrolled between July to December 2016. A control group included 20 healthy volunteers (informed consent was obtained).
Results: Patients and controls had similar sex and age. Median of JDM duration was 7(0-16) years and 5/20 (25%) had active disease, as defined by PRINTO criteria (2012). The median Myositis Damage Index (MDI) severity score was 0 (0-0.142) and the median MDI extent score was 0 (0-0.209). Conventional echo revealed preserved LV EF (≥ 55%) in all individuals, although values were lower in JDM (65 ±5.28%x75 ± 5.43%; $p = 0.005$). LV diastolic function parameters were similar. In the JDM group, 2DST identified significant reduction of LV peak systolic longitudinal strain (-18.2 ± 3.35%x-23.2 ± 3.45%; $p < 0.0001$) and strain rate (-1.1 ± 0.32 s-1x-1.35 ± 0.27 s-1; $p = 0.039$) and lower longitudinal strain rate in early diastole (1.63 ± 0.37s-1 × 1.97 ± 0.62 s-1; $p = 0.047$). LV peak systolic circumferential strain and strain rate and circumferential strain rate in early diastole were not different. There was a negative correlation between creatine-phosphokinase (CPK) levels and LV peak systolic longitudinal strain ($r = -0.5$; $p = 0.023$). Patients with a MDI severity score >0.027 showed lower LV peak systolic longitudinal strain than those with MDI ≤0.027 (-15.34 ± 3.3%x-19.14 ± 2.87%; $p = 0.024$).
Conclusions: LV longitudinal 2DST derived strain was able to detect early systolic and diastolic dysfunction in pediatric JDM patients. LV peak systolic longitudinal strain impairment seems to correlate with muscular damage and CPK levels.

P1780 - INTRACARDIAC ECHOGENIC FOCI REVISITED

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Background/Hypothesis: The clinical importance of intracardiac echogenic foci(ICEF)on prenatal ultrasound remains debatable amongst pediatric cardiologists and obstetricians. It has previously been discredited as a marker of triploidy and no clear association with congenital heart disease (CHD) has been established. Yet it continues to be a frequent reason for referral for fetal echocardiogram. The aim of this study was to assess the presence of CHD in patients with ICEF and to determine if it represents echocardiographic artifact. We hypothesized that there is no significant association between ICEF and CHD and that ICEF represents echocardiographic artifact.
Materials and Methods: The institutional fetal echocardiography database was retrospectively searched for reports containing the diagnosis of echogenic focus from November 2014-November 2016. Each study was evaluated by a single reviewer. The presence of ICEF in the 3 standard views analogous to the apical 4 chamber, parasternal short axis, and parasternal long axis views was recorded. In addition, the presence of CHD, pericardial effusion, and/or fetal arrhythmia for each study was documented. Statistical analysis was performed using cross-tabulation with chi-square analysis.
Results: 55 patients, yielding 145 studies were reviewed. 6 patients (10.9%) had CHD, which included VSD(n = 3), PA/VSD(n = 2), and vascular ring(n = 1). ICEF was noted in all 3 views in 33 studies(23%), in 2 views in 45 studies(31%), in 1 view in 22 studies (15%), and resolved in 45 studies (31%)(see table). Of those studies with ICEF noted in all 3 views, 19%(n = 6) were associated with CHD, indicating a significant association in this highly selective population ($p = 0.039$).
Conclusions: There is a higher incidence of CHD in fetuses with ICEF compared to the general population. The presence of ICEF was more likely to be identified in multiple views, suggesting it is unlikely echocardiographic artifact.

P1787 - CHARACTERIZATION OF CARDIAC TUMORS IN CHILDREN BY CARDIAC MAGNETIC RESONANCE

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Introduction: Cardiac tumors are rare in childhood, with a prevalence of 0.0017 to 0.28% in autopsy series. In most cases the diagnosis is established by echocardiography, however, the ability to define the extent and determine the type of tumor is limited. The aim of this study is to analyze the clinical utility of cardiac magnetic resonance (CMR) in patients referred for a probable diagnosis of cardiac tumor.
Methods: Retrospective study. All CMR performed between 2002 and 2016 with a suspected diagnosis of cardiac tumor in the Clinical Hospital of the Pontifical Catholic University of Chile were included. The tests were performed for tissue characterization with sequences T1 and T2-weighted, STIR and T1W with fat suppression before and after administration of gadolinium intravenously; multi-plane cine sequences including ventricular function study; phase contrast flow measurements, angio-resonance, perfusion studies and myocardial viability.
Results: A total of 1,091 CMR studies were performed. In 2.8% (31) the diagnosis was cardiac tumor. The median age at diagnosis was 3 years (range 4 days - 15 years). 55% were women. 32.3% were rhabdomyomas, 25.8% fibromas, 16.1% malignant, 12.9% vascular and 12.8% others (thrombi, papillary fibroelastoma and myxoma). There was 100% concordance between CMR and biopsies performed.
Conclusions: The findings in our series are similar to those described in the literature. CMR is very useful in the evaluation of cardiac tumors, because in the majority of the cases determines its etiology through tissue characterization. In addition, it accurately defines the location, extent, borders of the tumor and the functional impact either secondary to the involvement of cardiac structures or to obstructive phenomena.

P1789 - BIVENTRICULAR IMPACT OF MILD TO MODERATE PULMONARY VALVE STENOSIS IN FETAL LIFE

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Background: Pulmonary valve stenosis (PS) represents a common cardiac defect. There is limited information on fetal cardiovascular changes associated with PS. Such information may be valuable to improve prenatal counseling and postnatal management.
Methods: A comprehensive echocardiography including morphometric and functional parameters was performed on 16 fetuses with isolated PS and 48 controls matched by gestational age at scan (+ /- 2 weeks). Cases were follow-up until 12 months of life. Fetal PS-cases were grouped according to the need of postnatal pulmonary valvuloplasty (PV).
Results: The study groups were similar in fetal ultrasound and perinatal characteristics. Median gestational age at PS diagnosis was 33.4 weeks (range 20.0-36.5) and most cases corresponded to mild and moderate PS. Six newborns required performing a postnatal PV. Fetuses with PS presented with larger and more globular hearts with increased myocardial wall thickness (Figure 1). Despite preserved RV systolic function, PS-cases showed increased biventricular cardiac output (CO) and signs of diastolic dysfunction, with higher ductus venosus pulsatility index (0.72 ± 0.31 vs. 0.53 ± 0.16, $p < 0.004$) and tricuspid E/E´ratio (7.52 ± 3.07 vs 5.76 ± 1.79, $p < 0.02$). Fetuses with PS displayed a compensatory increase in left ventricular (LV) ejection fraction ($79.3 \pm 8.23\%$ vs. $67.6 \pm 11.3\%$, $p < 0.003$) and mitral annular-plane systolic excursion (5.94 ± 1.38 mm vs. 5.0 ± 1.22 mm, $p < 0.03$). Finally, fetuses requiring a postnatal PV showed a significantly smaller RV and

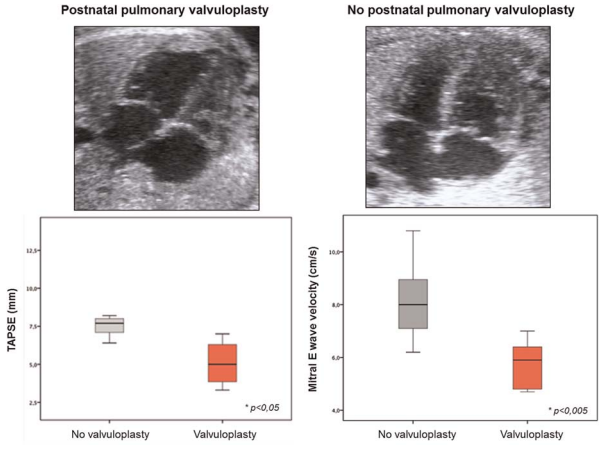

Figure.

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pulmonary valve diameter, reduced tricuspid annular-plane systolic excursion, increased LV CO and more pronounced signs of LV diastolic dysfunction.
Conclusions: fetuses with PS presented with more hypertrophic, larger, and globular hearts at third-trimester of pregnancy associated with a higher biventricular CO, impaired biventricular relaxation and signs of increased LV systolic function. Our data suggest that RV and LV functional parameters could be useful for predicting the need for.

P1795 - IDENTIFYING SICKLE CELL ANEMIA PATIENTS WITH PULMONARY HYPERTENSION USING 2D SPECKLE TRACKING STRAIN MEASUREMENTS

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Background: Sickle cell anemia (SCA) is a common hemoglobinopathy. Pulmonary hypertension (PH) is a known complication of SCA and can contribute to premature death. Continuous wave spectral Doppler interrogation of the tricuspid regurgitation (TR) jet is used to estimate pulmonary artery pressure (PAP). Limited echocardiographic acoustic windows or the absence of TR may preclude estimation of PAP. 2D speckle tracking right ventricle strain offers a unique tool to assess changes in ventricular performance and therefore may allow identification of patients with such changes secondary to pulmonary hypertension. The present study evaluates global longitudinal strain (GLS) imaging in SCA patients with and without pulmonary hypertension.
Methods: Thirty-eight patients with SCA were included in the study. Twenty-one patients with TR velocities greater than 2.5 m/s were compared to seventeen age-matched patients with normal estimated PAP. GLS of the right and left ventricles was assessed. Non-parametric analysis was performed using Mann Whitney U test.
Results: The group without pulmonary hypertension ($N = 17$) had a median RV strain of -16.2 and an interquartile range (IQR, or the 75th percentile – 25th percentile) of 4.70. The group with pulmonary hypertension ($N = 21$) had a median RV strain of -12.7 and IQR of 6.43. These median values were significantly different ($U = 75$, $p = 0.002$). There was no difference in LV strain values between the two groups.
Conclusions: Global RV strain values are significantly lower in SCA with PH compared to SCD patients with normal estimated pulmonary pressure. RV GLS can be performed during echocardiographic evaluation of SCA patients for PH and may have utility in identifying such patients when there is inadequate TR to estimate PAP. Ranges of RV and LV GLS in patients with hemoglobinopathies continue to be established.

P1806 - CARDIAC MAGNETIC RESONANCE IMAGING AND ECHOCARDIOGRAPHIC PREDICTORS OF SINGLE VERSUS BIVENTRICULAR REPAIR IN A LARGE COHORT OF PATIENTS WITH WELL FORMED VENTRICLES AND COMPLEX GREAT ARTERIAL RELATIONS

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Background: Decision-making regarding single (SV) or biventricular(BV) repair can be challenging. Areas of concern include valvular function and the potential route for baffling the proposed systemic ventricle to the aorta. We present a cohort of these patients, assessed by cardiac magnetic resonance (CMR) and echocardiography prior to decision-making.
Materials and Methods: Patients with complex ventriculoarterial connections, referred for CMR assessment (August 2005-October 2016) to aid in determining SV versus BV repair were identified. Demographics, procedures and clinical course were recorded, as well as volume and vascular data from CMR and echocardiogram closest to the time of CMR. MRI volumes were measured both with hypothetical baffle closure of the ventricular septal defect (VSD) as well as imaginary closure of the defect as a continuation of the septum.
Results: Forty-seven CMR scans(forty patients) were identified. Ten scans in 8 patients were excluded(1 death pre-definitive surgery, 7 surgery awaited). Twenty-one patients underwent SV palliation and 16 BV. There was no significant difference in the great vessel arrangement between the two groups, but heterotaxy syndrome was significantly associated with SV palliation (SV 47.4%, BV 14.3%, $p = 0.035$) as was non-committed VSD or a partial/complete canal defect (SV 47.5%, BV 7.1%, $p = 0.011$). VSD size was not significantly different. There were no significant differences between the groups in CMR volumes or changes in CMR volumes with virtual baffle, although the change in the right ventricular end-diastolic volume approached significance (SV -15%, BV -9%, $p = 0.085$).
Conclusions: There were no significant differences between patient groups in CMR volumetry, although patients with severely hypoplastic ventricles may have already been selected for a SV pathway. Presence of heterotaxy or a non-committed or canal-type defect were predictors of SV pathway. The size of the VSD was not significantly different, suggesting that position rather than size of the VSD is more important.

P1832 - INTRAVENTRICULAR KINETIC ENERGY IN HEALTHY CHILDREN AND IN THOSE WITH HYPOPLASTIC LEFT HEART SYNDROME

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Background: In hypoplastic left heart syndrome (HLHS) the right ventricle (RV) performs as a systemic ventricle, however the adaptation is incomplete. Patients experience early circulatory failure and those with the largest left ventricle (LV) remnants have poorer outcomes. Intra-cardiac kinetic energy (KE) differs between the healthy LV and RV and is sensitive to changes in systolic and diastolic function. We compared intra-cardiac KE in healthy children to those with HLHS to study the energetic limits imposed on the systemic RV and how residual LV size affects the KE profile.
Methods and Methods: 4D flow MRI was acquired in 8 healthy children (1-16 years) and 30 children with HLHS (1-16 years) undergoing clinical MRI (15 HemiFontan, 15 Fontan). Intraventricular KE was measured throughout the cardiac cycle and indexed to ventricular volume. LV remnant was grouped into shape (slit-like or globular) and size (median volume = 2.4mls).
Results: Two intra-cardiac KE peaks were present reflecting systole, and the fused inflow of early diastole. The healthy LV displayed a larger diastolic peak compared to the healthy RV ($p < 0.05$). Compared to the healthy RV those with HLHS had a similar diastolic peak in the presence of preserved function, but

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despite increased mass ($p < 0.001$) and volume ($p < 0.05$). No differences emerged between stages of surgery. Globular shaped LV remnants had the lowest RV diastolic KE compared to slit-like LV remnants ($p = 0.005$). This was also related to the size of the LV remnant with lower KE seen in those with larger median LV size (LV >2.4mls; $p = 0.016$).
Conclusions: In HLHS, the RV displays incomplete physiological adaptations to its role as a systemic ventricle with diastolic KE limited by the inherent RV morphology. This is further influenced by larger, globular LV remnants acting as stiff piggy-back ventricles with adverse effects on diastolic momentum.

P1864 - INITIAL EXPERIENCE IN THE USE OF CARDIAC MAGNETIC RESONANCE IN THE EVALUATION OF THE BORDERLINE LEFT VENTRICLE

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Introduction: The definition of left ventricular (LV) hypoplasia is based on the presence of a left ventricle unable to sustain the systemic circulation. Intermediate forms, called the borderline LV, represent a challenge during the neonatal period because its evaluation will determine the biventricular or univentricular repair, with important implications for the prognosis. As a complementary study, the use of cardiac magnetic resonance (CMR) has been increasing due to its ability to provide valuable information.
Objective: To show the initial experience in the use of CMR as an additional tool in the evaluation of borderline LV.
Methods: All patients evaluated at the Clinical Hospital of the Pontifical Catholic University of Chile between 2013 and 2016, with a LV volume measured by echocardiogram of less than 20 mL/mt2 in which there was controversy about the surgical approach were included. All patients underwent an echocardiogram and CMR during the first month of life, prior to surgery.
Results: 5 patients were recruited, all female. Follow-up was from 5 to 27 months. Echocardiography underestimated LV volume compared to the CMR measurement, with differences between 10.1 and 23.5 mL/mt2. In one case, the LV volume was less than 20 mL/mt2 and the flow through the LV outflow tract was less than 1 L/min/mt2. This patient underwent univentricular repair and the other four cases followed the biventricular route. Four patients underwent interventional catheterization (3 patients who followed the biventricular pathway and the patient who followed the univentricular approach). These interventions were not related to the choice of the surgical pathway. No deaths were recorded during follow-up.
Conclusions: CMR is a valuable complementary tool in the assessment of borderline LV. Studies with a greater number of patients and long-term follow-up are needed.

P1868 - PSEUDOANEURYSM OF THE LEFT VENTRICULAR OUTFLOW TRACT IN AN INFANT

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Introduction: Pseudoaneurysm of the left ventricular outflow tract (LVOT) is rare but potentially fatal. It may develop following infection, cardiac surgery, trauma or systemic inflammation. We describe a case of LVOT pseudoaneurysm in an infant with complex congenital heart disease and multiple previous surgeries.
Case Report: An 11-month-old boy with repaired double outlet right ventricle (DORV), transposed great arteries (TGA) and large inlet ventricular septal defect (VSD) presented with three months of wheeze. Previous surgery included Arterial Switch with PA band placement in the neonatal period. At 6 months of age, he underwent VSD closure and creation of a LV-aorta tunnel, including resection of a large conus. The onset of wheeze was associated with fever and coryza, however it persisted beyond the infective symptoms. Echocardiogram demonstrated a large aneurysm between the anterior mitral valve leaflet and aortic annulus. Cardiac CT angiogram confirmed the presence of a large, lobulated aneurysm (40x28 × 22mm) arising from the lateral aspect of the LVOT and projecting posterosuperiorly. Mass effect on the left atrial appendage, left common pulmonary vein and distal left main bronchus with gas trapping was apparent. At operation, a large false aneurysm of the LVOT extending posterior to the aorta and cephalad to the left atrium was identified. The walls were thick and muscular with a neck measuring 12mm, originating from the site of previous surgery. It was hypothesised that previous resection of the conus had weakened the mitral-aorta continuity. Full resection was achieved by exposing the false aneurysm from the aortic and mitral valves, and reconstructing the LVOT with CardioCel. Twelve months post-operatively, the child remains asymptomatic with no residual aneurysm and a widely patent LVOT on echocardiogram.
Conclusion: Pseudoaneurysms of the LVOT are rare in children, with potentially devastating outcomes. Expedient surgical excision is required due to the risk of rupture.

P1879 - THE DESCENDING AORTIC DOPPLER IN VASCULAR RINGS PREDICTS PRESENCE OF DIVERTICULUM OF KOMMERELL

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Background: Vascular rings (VR) are rare congenital anomalies of the great arteries. We noted some patients with VR had descending aortic Doppler tracings similar to patients with coarctation of the aorta. This has not been described in the literature. We hypothesized that the descending aortic flow pattern in VR is different from normal, even without aortic obstruction, and correlates with anatomical characteristics of the VR.
Methods: We reviewed descending aortic Doppler features of pediatric and adult patients with different types of VR who were evaluated at our center from 2005 to 2016 and compared them with age-matched controls. Patients with a patent ductus arteriosus and left sided obstructive lesions were excluded. The anatomy of the ring and presence of diverticulum of Kommerell (DK) in patients with a right aortic arch and aberrant left subclavian (RAA) were confirmed by magnetic resonance imaging, computed tomography scan or operative data. Mann-Whitney, logistic regression and ROC analyses were performed.
Results: Of 80 patients, 28 (35%) had double aortic arch (DAA), 45 (56%) had RAA, of which 21 (47%) had confirmed DK. VR patients had higher end diastolic velocity and lower systolic/diastolic ratio compared to controls. This remained true when both RAA and DAA were each analyzed against controls (table 1). RAA patients with DK had lower pulsatility indices compared to those without a

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Diverticulum (p = 0.039) (table 2). Pulsatility index ≤4.12 was associated with DK with 62% sensitivity and 92% specificity.
Conclusions: The presence of elevated end diastolic velocity and lower systolic to diastolic ratio in the descending aortic Doppler of patients without arch obstruction suggests the possibility of a vascular ring while a lower pulsatility index in patients with RAA suggests the presence of DK. These findings may permit new insights into the aortic flow disturbances in these patients.

Table 1. Doppler variables in vascular ring patients compared to normal controls; RAA and DAA compared to normal controls.

Doppler Variable	VR Median (IQ range)	Control Median (IQ range)	P-value*	P-value **	All RAA Median (IQ range)
PSV(m/s)	0.83 (0.40)	0.85 (0.45)	0.307		0.86 (0.43)
EDV (m/s)	0.14 (0.07)	0.11 (0.06)	**<0.001**	**0.043**	0.14 (0.08)
S/D	5.74 (2.96)	7.05 (2.83)	**<0.001**	**0.007**	5.82 (3.19)
PI	5.26 (2.41)	5.65 (1.93)	**0.05**	0.448	4.97 (2.80)
AS (m/s)	0.77 (0.35)	0.81 (0.44)	0.31		0.81 (0.35)
DS (m/s)	0.78 (0.39)	0.83 (0.44)	0.260		0.82 (0.35)

Doppler Variable	Control Median (IQ range)	P-value *	All DAA Median (IQ range)	Control Median (IQ Range)	P-value*
PSV(m/s)	0.89 (0.48)	0.196	0.84 (0.40)	0.83 (0.46)	0.918
EDV(m/s)	0.11 (0.07)	**0.002**	0.14 (0.08)	0.11 (0.04)	**0.003**
S/D	7.20 (2.68)	**<0.001**	5.74 (3.13)	6.72 (2.82)	**0.004**
PI	5.41 (2.11)	0.064	5.56 (2.22)	6.02 (1.86)	0.339
AS (m/s)	0.87 (0.47)	0.143	0.78 (0.38)	0.77 (0.44)	0.727
DS (m/s)	0.87 (0.47)	0.102	0.81 (0.38)	0.79 (0.38)	0.658

Kommerell; PSV: Peak systolic velocity; EDV: End diastolic velocity; S/D: Systolic to diastolic ratio; PI: Pulsatility index [(maximum velocity - minimum velocity)/mean velocity], AS: Acceleration slope; DS: Deceleration slope; * Univariate p-value, ** Multivariate p value

Table 2. Doppler variables according to vascular ring subtypes and between subtypes compared to normal controls.

Doppler Variable	RAA - DK Median (IQ range)	Control Median (IQ range)	P-value*	RAA + DK Median (IQ range)
PSV (m/s)	0.87 (0.38)	0.89 (0.49)	0.716	0.80 (0.41)
EDV (m/s)	0.15 (0.08)	0.12 (0.04)	0.083	0.17 (0.11)
S/D	5.74 (3.33)	6.49 (2.91)	**0.019**	4.53 (3.025)
PI	5.63 (2.00)	5.62 (2.17)	0.769	3.77 (1.969)
AS (m/s)	0.84 (0.33)	0.88 (0.48)	0.634	0.74 (0.40)
DS (m/s)	0.83 (0.32)	0.87 (0.48)	0.597	0.77 (0.41)

Doppler Variable	Control Median (IQ range)	P-value*	RAA – DK v RAA + DK Median difference	P value*	P-value**
PSV (m/s)	0.90 (0.47)	0.136	0.07	0.40	
EDV (m/s)	0.13 (0.07)	**0.007**	0.02	0.06	0.73
S/D	6.86 (2.198)	**0.002**	1.21	0.03	
PI	5.08 (1.56)	**0.018**	1.86	**0.004**	**0.04**
AS (m/s)	0.87 (0.47)	0.113	0.10	0.48	
DS (m/s)	0.88 (0.47)	0.079	0.06	0.38	

VR: Vascular ring; RAA: Right aortic arch with aberrant left subclavian artery; DAA: Double aortic arch; DK: Diverticulum of Kommerell; PSV: Peak systolic velocity; EDV: End diastolic velocity; S/D: Systolic to diastolic ratio; PI: Pulsatility index [(maximum velocity - minimum velocity)/mean velocity], AS: Acceleration slope; DS: Deceleration slope; * Univariate p-value, ** Multivariate p value

P1900 - ECOCARDIOGRAPHIC EVALUATION OF PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM AND CRITICAL PULMONARY STENOSIS

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Background: To identify the echocardiographic criteria that guided the therapeutic behavior in pulmonary atresia with intact ventricular septum (PAIVS) and critical pulmonary stenosis (CPS) in newly born.
Materials: Descriptive and retrospective analysis based on the review of clinical records of 32 patients with PAIVS or CPS in newborn from 2008 to 2016 at Pedro de Elizalde Hospital. Patients were grouped according to the degree of RV development in: Mild hipoplasia (MI): TV Z score between -1 to > -2.5; RV > 30 cm3/m2; TV/MV ratio >0.8; PA Ring >1.25 cm/m2. Moderate (MO): TV Z score: -2.5 to -4.5; RV < 30 cm3/m2 and >10 cm3/m2; TV/MV: 0.5 < 0.8. Severe (SV): Z TV <-4.5; RV < 10 cm3/m2; ratio TV/MV <0.5; RV with two or fewer portions; Presence of coronary fistulas and coronary circulation RV dependent.
Results: Z score of the TV ring defined the initial behavior in 89% of patients (p). The results were: Biventricular: in 18/32p: 16p CPS and 2p PAIVS, RVOT permeabilization: Initial valvuloplasty in all MI (surgical valvotomy 3p MO hipoplasia, 2p shunts). Univentricular 12/32: 2p MO and 10p SV; 2 ended up as 1-1/2 ventricular. 6p has more than one coronary fistula; receive initially shunt. 3p deaths.
Conclusions: In PAIVS and CPS, the uni or biventricular future depends on several factors: type of atresia, RV anatomy, function, size of the tricuspid ring, amount of coronary fistulas in connection with RV and presence of RV-dependent coronary circulation. In our experience, the use of the TV Z score was the most reliable selection criteria, in the absence of significant tricuspid regurgitation or Ebstein anomaly, in which the TV/MV ratio and RV/LV volume was used as a more effective criterion for assessing the degree of development of the RV. If RV-dependent coronary circulation is present, no decompression of the RVOT will be indicated.

Table 1. Z score of pulmonary ring and initial tricuspid valve and last echocardiographic control in severe hypoplasia.

UNIVENTRICULAR	X	DS	P
Initial Z TV	-4,61	0,81	
Z TV latest control	-4,67	1,21	NS
Initial 2 PA	-3	0,71	
Z PA latest control	-3,67	1,7	0.000

Table 2. Z score of pulmonary ring and initial tricuspid valve and last echocardiographic control in mild hypoplasia.

BIVENTRICULAR	X	DS	P
Initial Z TV	-0,75	1,77	
Z TV latest control	1,50	0,71	0.05
Initial Z PA	-1,20	2,14	
Z PA latest control	1,07	1,62	0.02

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Table 3. Absolute value in centimeters of pulmonary ring and initial tricuspid valve and last echocardiographic control in mild hypoplasia.

BIVENTRICULAR	X	DS	P
Initial TV (cm)	1,08	0,17	
TV latest control (cm)	1,93	0,36	0.000
Initial PA (cm)	0.68	0,14	
PA latest control (cm)	1,21	0,43	0.017

Table 4. Characteristics of the population studied

Variable	Mild Hipoplasia (p = 15)	Moderate Hipoplasia (p = 5)	Severe Hipoplasia (p = 12)	Total (p = 32)
Male gender	7 (46%)	2 (40%)	8 (66%)	17 (53%)
Perinatal diagnosis	2 (13%)	–	4 (33%)	6 (18%)
TV Z score	-0.75 ±1,77 (0/-2)	-2,83 ±1,04 (-2/-4)	-4,61 ±0,81 (-4/-6)	
PA Z score	-0,93 ±0,86 (-0,75/-2,5)	-1,33 ±1,15 (0/-2)	-3,67 ±1,89 (-1/-6)	
Sinusoidcs en VD	–	1 (20%)	6 (50%)	7 (22%)
RV tocnderji CC	–	–	3 (25%)	3 (9%)
Ebstein anomaly	–	1 (20%)	–	1 (3%)
Severe tricuspid regurgitation	–	3 (60%)	2(17%)	5 (16%)

P: patient, TV: tricuspid valve, MV: mitrall valve, RV: Right ventricle, CC: coronary circulation, PA: pulmonary artery.

P1902 - EXTENSIVE MYOCARDIAL CALCIFICATION IN A NEWBORN INFANT DETECTED BY ECHOCARDIOGRAPHY IN ENTEROVIRUS MYOCARDITIS

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A 7 day old female was admitted to ICU with persistent junctional re-entry tachycardia. She was subsequently diagnosed with an enterovirus (Coxsackie B4) causing myocarditis and poor systolic function requiring VA ECMO. Large echogenic masses in the myocardium were observed on echocardiography one week after presentation. The patient remained in heart failure and developed echocardiographic features of severe restrictive cardiomyopathy. The family declined more intensive treatment and the baby died aged 61 days. This is the first time we have seen this echocardiographic picture. A full review of the chart was performed including all clinical investigations and imaging. The autopsy report and histology were reviewed and discussed with the histopathologist. In addition a literature search was performed to review myocardial calcification and myocarditis in the newborn. The initial echocardiogram showed normal cardiac anatomy, reduced ventricular systolic function and normal myocardial appearance. Echogenic masses were noted in the interventricular septum (IVS) on Day 10. Serial echocardiography demonstrated deteriorating systolic and diastolic ventricular function, worsening AV valve regurgitation and diffuse effusions. The masses became highly echogenic and some were also detected in the left ventricular (LV) free wall. On autopsy, extensive damage due to myocarditis with marked calcification, fibrosis and persistent chronic inflammation was documented. It was noted to be particularly marked in the IVS and LV free wall with damage to 30-40% of the cardiac mass. This resulted in a restrictive cardiomyopathy causing congestive cardiac failure and widespread effusions and oedema. These features were consistent with those demonstrated by echocardiography. We described an unusual presentation of early myocardial calcification of a newborn with severe myocarditis. There is a recognised association of Coxsackie B with dystrophic myocardial calcifications. Myocardial inflammation, fibrosis and necrosis lead to calcium phosphate deposits. This is an indication of severe, usually irreversible myocardial damage and predicts a poor outcome.

P1919 - PEDIATRIC ECHOCARDIOGRAPHIC NOMOGRAMS FOR TWO DIMENSIONAL RIGHT VENTRICULAR SUB COSTAL VIEW DIAMETERS AND AREA A NEW ADDITIONAL APPROACH TO MEASURE THE RIGHT VENTRICLE

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Background: The sub-costal examination is part of routine examination in pediatric echocardiography, and may allow to visualize right ventricular (RV) dimensions and function offering the advantage to include also the infundibulum. Despite this nomograms for RV dimension in sub-costal view are lacking.
Methods: We prospectively studied healthy Caucasian Italian children (0-18 yrs) by two-dimensional echocardiography. Echocardiographic measurements included: sub-costal end diastolic and end-systolic area (subc-RVED-area, subc-RVES-area) and basal-apical and later-lateral (at the tricuspid valve attachment) diameters (subc-RVED-AP-diameter, subc-RVED-ll-diameter, subc-RVES-AP-diameter, subc-RVES-ll-diameter), 4 chamber end diastolic and end systolic area (4ch-RVED-area, 4ch-RVES-area) and length (4ch-RVEDl, 4ch-RVESL) end diastolic basal and mid-cavity diameters. Heteroscedasticity was accounted for by White or Breusch-Pagan test. Age, weight, height, heart rate and body surface area (BSA) were used as independent variables in different analyses to predict the mean values of each measurement. Structured Z scores were then computed. Agreement analysis of RV diameters and areas in sub-costal and 4-chamber view were made.
Results: In all, 732 subjects (age 0 days-17 years; 48% female; BSA 0.12-2.12 m2) were studied. The Haycock formula was used when presenting data as predicted values (mean ± 2 SDs) for a given BSA and within equations relating echocardiographic measurements to BSA. The predicted values and Z-score boundaries for all measurements have been presented. Good correlations were found among diameters and area calculated in sub-costal view with those evaluated in 4-chamber view. Pearson's coefficients were: Subc-RVED-AP-diameter vs 4-chRVED-length ($r = 0.84, p = 0.000$), subc-RVES-AP-diameter vs 4-chRVES-length ($r = 0.69$, $p = 0.000$), subc-RVED area vs 4ch-RVED-area ($r = 0.872$, $p = 0.000$), subc-RVES area vs 4ch-RVES-area ($r = 0.784$, $p = 0.000$).
Conclusions: We report pediatric echocardiographic nomograms for RV diameters and areas calculated by sub-costal view. Good correlations emerged among RV measurements in sub-costal view and those obtained in 4-chamber view.Our data may implement normative data for 2-dimensional echocardiography evaluation of the RV in children.

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Table 1. Coefficients for regression equations relating echocardiographic measurements end body surface area, the Standard Etr or of the Estimate, the determination coefficient. Normality test: Shapiro-Wilk and Lilliefors (Kolmogorov-Smimov). Heteroscedasticity test (White test and Breusch pagan test). BSA HAYCOCK (ln[y] = a + b*ln[x]); Z value = (in[Measurement]-(intercept + B*ln[BSA]))/√MSE

Measurement	Intercept	B	SEE (√MSE)	R^2	SW	KS	BP	W
Sabc-RVED-AP-diometer (mm)	4.161	0.538	0.095	0.878	0.057	0.182	0.884	0.723
Stibc-RVED-LL-diameter (mm)	3.189	0.488	0.145	0.716	0.275	0.088	0.398	0.525
Subc-RVED-area (mm)	2.723	1.002	0.190	0.861	0.138	0.200	0.461	0.557
Subc-RVES-AP-diameter (mm)	3.477	0.535	0.129	0.770	0.018	0.200	0.367	0.051
Subc-RVES-LL-diameter (mm)	2.918	0.539	0.159	0.692	0.002	0.009	0.800	0.143
Subc-RVES-ana (mm)	1.658	1.106	0.222	0.834	0.222	0.200	0.266	0.001

P1929 - ISOLATED MIDDLE LOBE SCIMITAR VEIN TO RA WITH VANISHING RIGHT MIDDLE PULMONARY VEIN

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Background: Scimitar syndrome comprises of pulmonary sequestration with total or partial drainage of right lung mainly to IVC along with aberrant supply from descending aorta. We are presenting rare case of isolated middle lobe scimitar to RA with normal drainage of right lower lobe vein. Right upper vein has got dual supply to LA via small caliber vein and through scimitar vein via intercommunicating channel.Test Balloon occlusion of scimitar vein demonstrated the presence of native hypoplastic middle pulmonary vein which was not demonstrated during initial catheterization and CT scan.

Description: 12 years old girl referred for dyspnea class II. Examination revealed wide and fixed split S2 with mid systolic ejection murmur at second intercostal space. Echocardiographic evaluation revealed large ostium secundum ASD with adequate rims and normal pulmonary venous drainage. While attempting ASD device closure it was realised that right middle pulmonary vein with scimitar morphology was draining abnormally at SVC to RA junction. Right upper lobe lung having dual supply via scimitar vein to RA through intercommunicating channels as well to LA through hypo plastic right vein. Right lower pulmonary vein was draining normally to LA. Test Balloon occlusion of scimitar vein with 16 × 40 TYSHAK balloon revealed hypoplastic right middle vein to LA which was not seen earlier. In view of hypoplastic right middle and upper pulmonary veins and hypo plastic right middle lobe option of rerouting of pulmonary veins by blocking scimitar vein was not considered. Patient underwent successful surgical ASD closure and rerouting of scimitar vein to left atrium.

Conclusion: Typical profile of the middle pulmonary vein draining into RA with associated absent middle lobe bronchus clinches rare diagnosis of isolated middle lobe scimitar. Redirecting blood flow through upper or middle lobe veins through intercommunicating channels is possible if caliber of

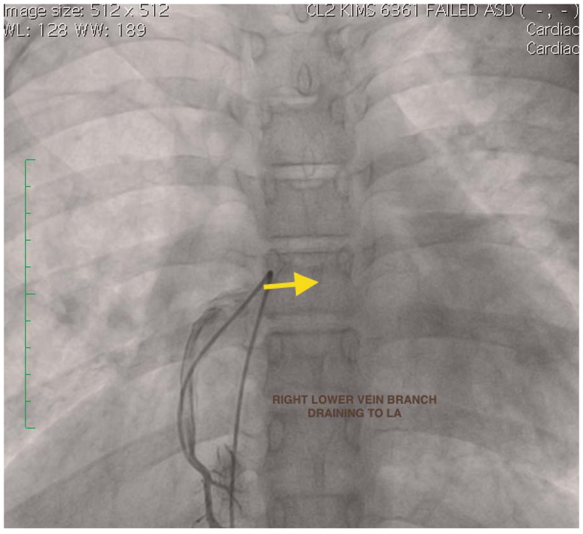

Figure 1.

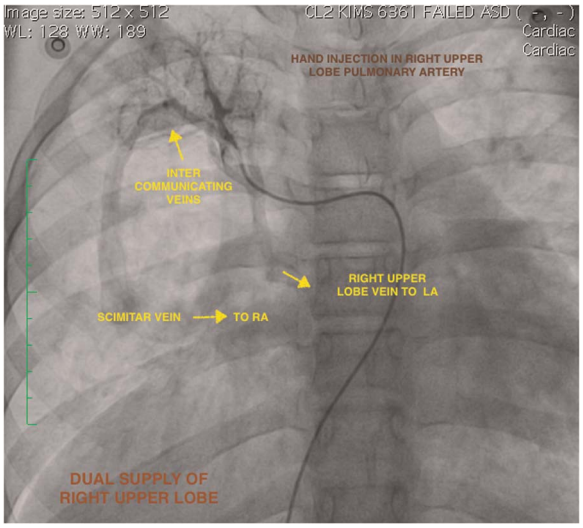

Figure 2.

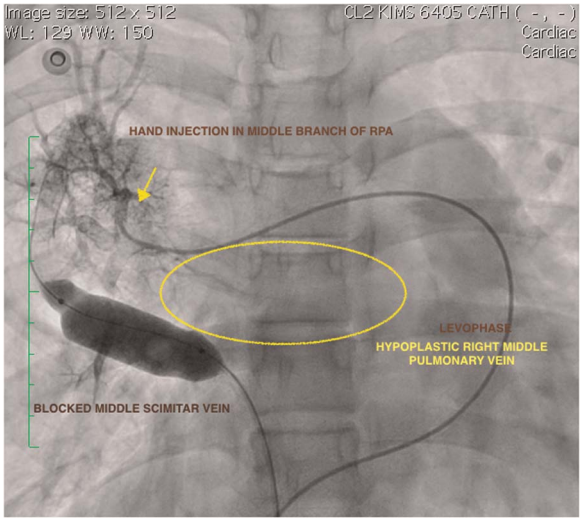

Figure 3.

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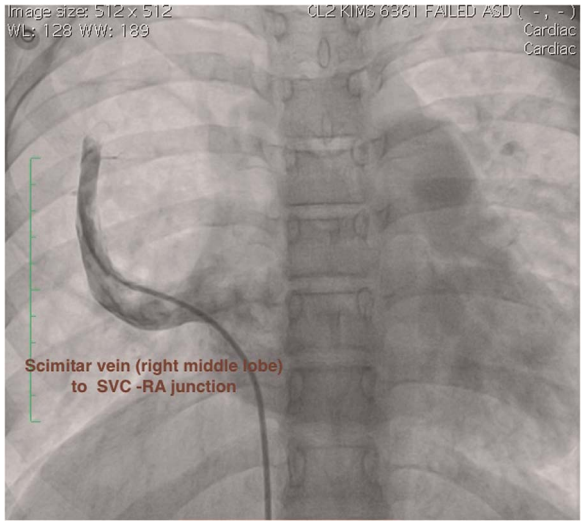
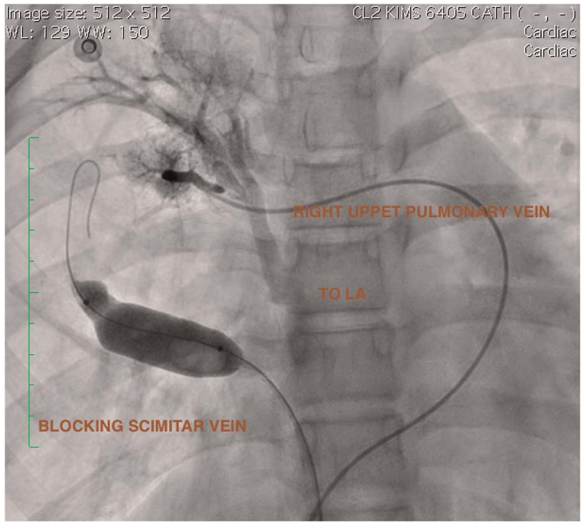


Figure 4.



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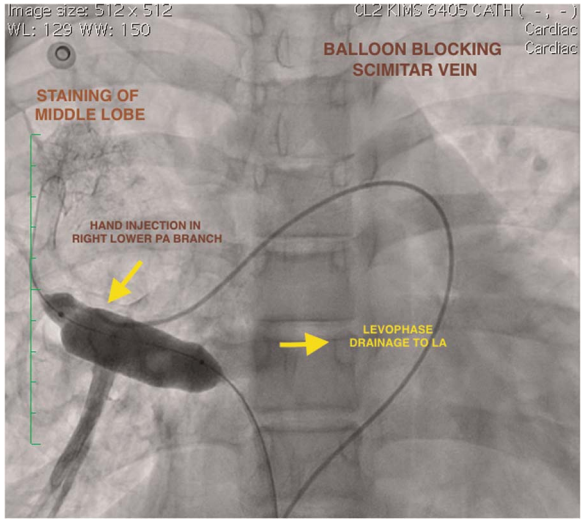

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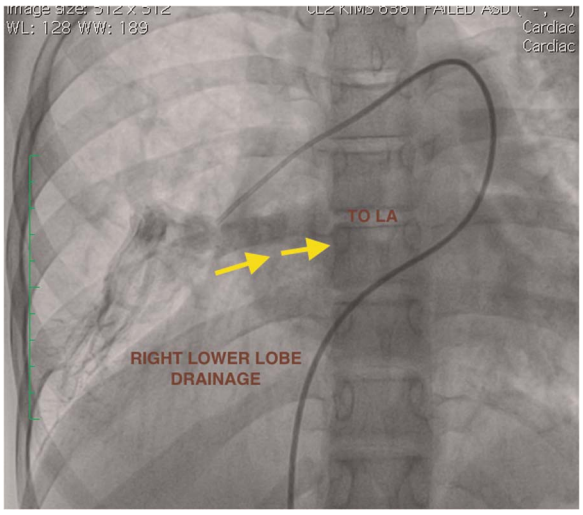

Figure 7.

P1936 - ISOLATED CONNECTION OF A LEFT SIDED SUPERIOR VENA CAVA TO THE LEFT ATRIUM A RARE CAUSE FOR CENTRAL CYANOSIS

Michael Kaestner[1], *Ronald Handke*[2], *Olchvary Julia*[3], *Pfeifer Jochen*[3], *Rentzsch Axel*[3], *Abdul-Khaliq Hashim*[3]
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Background: Congenital heart disease is a well-known cause for cyanosis in children which usually resolves after surgical correction of the respective defect. However, not every abnormality needs surgical correction but may have an impact for the patient when in need for medical treatment. We report a rare case of isolated connection of a left sided superior vena cava (SVC) to the left atrium and the clinical consequences.
Case: A 7y/o boy was incidentally found to have low oxygen saturation measured on an open house presentation of the local ambulance service. This was confirmed by repeated measurements by a pediatric cardiologist, who suspected abnormal drainage of a persisting left superior vena cava on echocardiogram. Magnetic resonance imaging demonstrated this finding with an isolated connection of a left superior vena cava to the left atrium (Figure). There was no connecting vein to the right SVC.
Conclusion: The parents refused surgery and the boy was send home with an emergency health card for documentation of this finding as this has an impact in case of possible therapeutic intervention and medical treatment.

1. Any kind of intravenous line should not be done from the left arm or left neck side or all medication will be applied to the arterial system directly and the patient is at high risk for cerebral and arterial complications.
2. The electrodes of electric devices like pacemakers or automatic implantable cardioverter defibrillator should be implanted from the right veins or need surgical implantation with epicardial electrodes.
3. Open heart surgery on heart-lung-machine will need selective cannulation of the left SVC.
4. In case of cardiac transplantation, the surgeon should do end to side connection of the left SVC to the right SVC or should perform left sided bidirectional cavo-pulmonary anastomosis.

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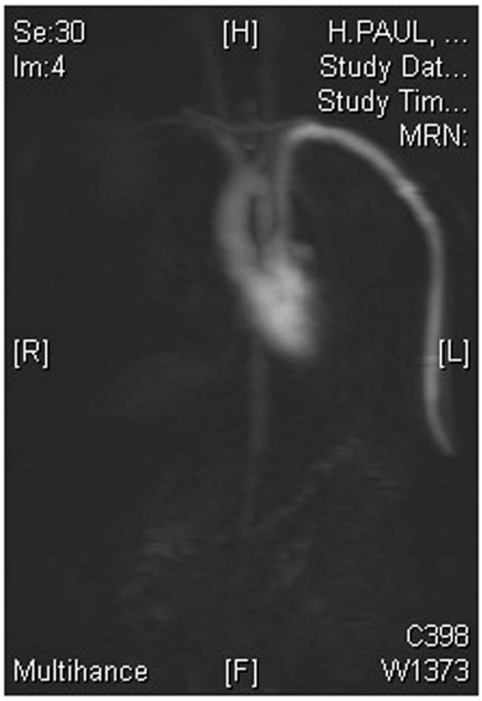

Figure.

P1943 - MYOCARDIAL DEFORMATION IMAGING FOR ASSESSMENT OF YOUNG PATIENTS WITH KAWASAKI DISEASE

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National Heart Hospital, Department of Paediatric Cardiology, Sofia-Bulgaria[1]; National Heart Hospital, Department of Cardiology, Sofia-Bulgaria[2]; National Heart Hospital, Department of Paediatric Cardiac Intensive Care, Sofia-Bulgaria[3]; University Hospital, Clinic of Paediatrics, Sofia-Bulgaria[4]; Tzaritza Ioanna Hospital, Department of Paediatric Oncology and Haematology, Sofia-Bulgaria[5]

Aim: To assess global and regional left ventricular (LV) function and to predict the long-term prognosis of Kawasaki disease patients.
Material and Methods: 82 children were included in the study – 62 with Kawasaki disease(mean age 9,4 ±3,9 years), 8 of whom with a residual coronary aneurism and 20 healthy controls (mean age 9,6 ±4, 8). Apical and basal short axis for 2D images were acquired in addition to apical four, three and two-chamber views. Global and regional peak systolic strain - longitudinal, radial and circumferential on the LV was derived and the strain curves (eLL, eCC, eRR) were extracted using a commercial software. Data are presented as means ± standard deviation. Student's T-test integrated in SPSS 19.0 was used. A value of $p \leq 0.05$ was considered significant.
Results: LV function was normal in both Kawasaki disease with no aneurysms and healthy controls groups, but there was a significat difference in the ejection fraction 65,39 ± 4,1vs. 71,9 ± 3,2 ($p = 0.003$), LV diastolic diameter (LVDD) z-score 0.7 ± 1,0 vs.-0,3 ± 0,8 ($p = 0.001$) and global longitudinal strain(GLS) measurements of the LV -19,23 ± 1,98% vs -21,73 ± 0.92% ($p = 0.03$). Significantly reduced GLS was found in the group of Kawasaki patients with aneurysms compared to controls -17,94 ± 1,69% vs -21,73 ± 0.92%. ($p < 0{,}001$) The regional peak systolic longitudinal strain (LS) was also reduced significantly, especially on the basal and middle LV – segments.
Conclusion: Decreased segmental regional LS values are a sign of regional hypokinesia with a possible local segmental ischemia in the group of Kawasaki with coronary aneurysms and are certain predictor of development of LV dysfunction in the future. Lack of significant differences of GLS in the Kawasaki group without an aneurysm is associated with still normal or lightly impaired global LV function and these group shoud be closely observed for ventricular dysfunction in the future.

P1952 - LIMITATIONS AND POTENTIAL OF CLINICAL APPLICATION USING PATIENT SPECIFIC THREE DIMENSIONAL HEART MODELS THREE YEAR EXPERIENCE OF SIMULATION OF CARDIAC SURGERY AND CATHETER INTERVENTION

Koichi Kataoka[1], Masaaki Kawada[2], Daisuke Matsubara[3], Kensuke Oka[3], Shun Suzuki[3], Takaomi Minami[3], Tomoyuki Satoh[3]
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Background and Objective: In 2014, we established simple and inexpensive methods using a personal three-dimensional (3D) printer to create patient-specific 3D heart models for planning of surgery, simulation of catheter intervention, explanation for patients/their families, and education of trainees/medical students. Here we report the limitations and potential of clinical application using 3D models through our 3-year experience.
Methods: Using OsiriX (Pixmeo, Switzerland), DICOM (Digital Imaging and Communications in Medicine) data of contrast-enhanced MDCT (multi-detector computed tomography) scan were converted into 3D image data and stored as STL (Standard Triangulated Language) files for 3D printing. UP Plus 2 personal 3D printer (Tiertime Technology, People's Republic of China) was subsequently utilized to print ABS (acrylonitrile butadiene styrene) resin solid model (solid model) for each lesion. If necessary, transparent silicone hollow model (hollow model) was additionally created using solid model as a mold. All processes were performed at our own institution.
Results: The time/cost needed to create solid and hollow models were 5–24 h/5,000 JPY and 3–7 days/10,000–20,000 JPY, respectively. Solid model was helpful in understanding the spatial relationship among the great vessels and trachea and so on. Hollow model had good reproducibility for understanding intracardiac structures and was useful in the surgical planning of complex/rare anomalies, even if their vascular wall thicknesses were not very accurate. Although its hardness rendered it unsuitable for training in detailed surgical techniques, hollow model was suitable for simulation of occluder deployment for patent ductus arteriosus with calcification in adults. X-ray transparency of hollow model was appropriate for catheter manipulation; because the catheter/device was visible from the outside, it was useful for training in catheter interventions.
Conclusions: From the viewpoint of fidelity for anatomical accuracy, there were some limitations in these models; however, high-quality simulation becomes possible using these features.

P1954 - A NOVEL FIBRILLIN 1 GENE MISSENSE MUTATION ASSOCIATED WITH MARFAN SYNDROME A CASE REPORT

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Background: Marfan syndrome (MFS) has highly variable clinical manifestations, resulting from genetic heterogeneity of pathogenic variants of the fibrillin-1 gene (FBN1) and TGFBR2 gene. We report a novel mutation of FBN1 gene in a case with MFS, who presenting rather Ehlers-Danlos syndrome like phenotypic manifestations

Case: A two-year-old girl was referred to the hospital with complaints of cardiac murmur and hypermobile joints. She was observed to have a dolichocephaly with down-slanting palpebral fissures, loose skin, and skeletal abnormalities including long and thin fingers, marked joints laxability, pectus deformity, scoliosis and pes planus. Her height was 50 percentile for age and the arm span to height ratio was 1.0. The grade 2 to 3 precordium regurgitant murmur was noted on apex. The severe farsightness and depigmentation of retinal pigment epithelium in fundus were identified, but no ectopia lentis on ophthalmologic examination. The progressive severe mitral valve insufficiency, aortic root ectasia (z score 2.8-3) and enlarged LVID (z score >2) were revealed on echocardiographic examination (Fig. 2). The chromosome analysis showed normal karyotype (46,XX). The genetic testing of COL5A1, COL5A2 gene for Ehlers-Danlos syndrome was normal. A novel missense mutation of FBN1 gene was identified with diagnostic exome sequencing. A heterozygous base substitution, c.3222 C > G in exon 27, resulting in a cysteine substitution by tryptophan at codon 1074 (p.1074 C > W) (Fig. 3) . No mutation was found in the TGFBR2 gene. The diagnosis was established by aortic root ectasia (z >2) and a bona fide FBN1 mutation. Her systemic scoring by revised Ghent nosology was 12 at five year od. Pedigree revealed her estranged father and a paternal cousin who apparently share marked joints laxability similar to that of proband. She was treated with beta-blocker and Angiotensin receptor blocker.

Conclusion: FBN1-associated fibrillinopathies are a group of diseases with dynamic phenotypic correlations.

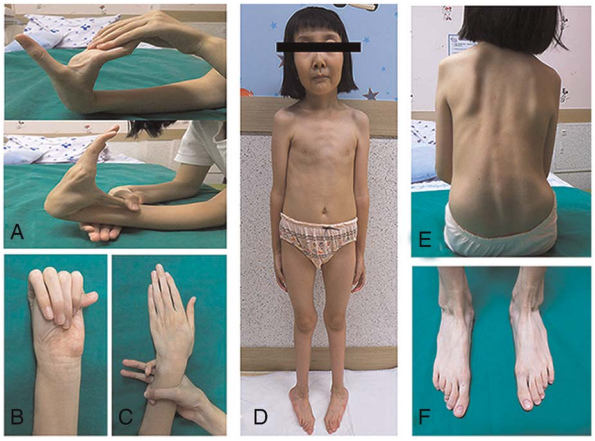

Figure 1.

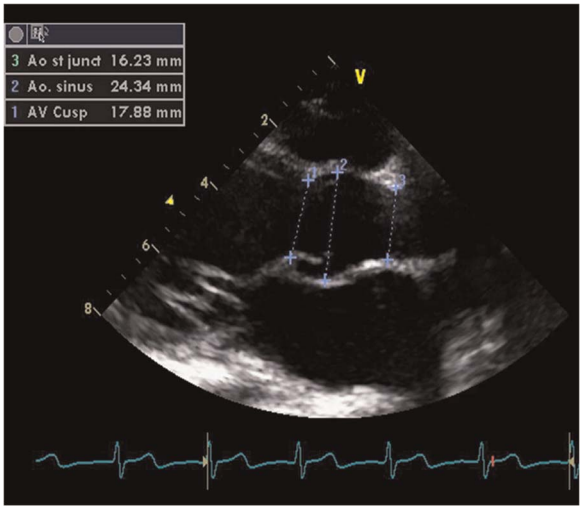

Figure 2.

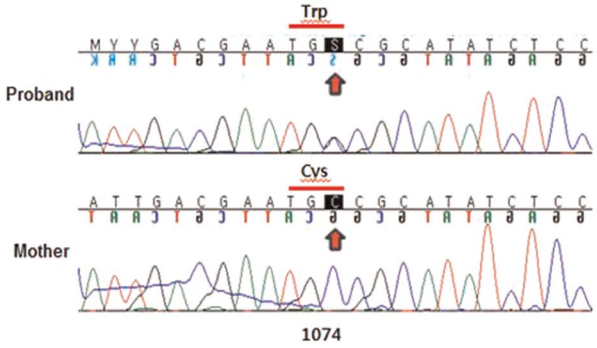

Figure 3.

P1974 - SUSTAINED SUBMAXIMAL ISOMETRIC HANDGRIP AS HEMODYNAMIC STRESSOR AFTER COARCTATION REPAIR IN CHILDREN

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Introduction: Patients with repaired coarctation remain at higher risk of hypertension and LV dysfunction despite successful repair at young age and abnormalities may initially be present only when the heart is stressed, i.e. during exercise. Echocardiography is difficult or impossible to do during a bicycle exercise test. This is the first study where submaximal isometric handgrip test is studied in conjunction with echocardiography in children with repaired coarctation.

Methods: Case-control study with 19 children with repaired coarctation (mean age 12,78y ± 2,33) compared to 19 matched controls. Isometric handgrip test with echocardiography followed by bicycle ergometry was performed in both groups.

Results: During exercise the blood pressure increased from 114 ± 11/64 ± 4 mmHg to 132 ± 14/79 ± 7 mmHg. There was no significant difference found between the groups. HR increased from 76 ± 22 bpm to 88 ± 21 bpm. ΔHR was significantly lower in coarctation patients compared to controls (p = 0,01). LVM was

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106 grams with no difference between the two study groups. All subjects had normal FS (>28%). E decreased with 2,73%, A increased with 20,33% whereas E/A decreased with 25,46% during exercise. E and A wave were significantly prolonged in coarctation patients at rest and during isometric exercise ($p < 0,05$). Tissue Doppler imaging of the ventricles: E' decreased with 3,88%, A' increased with 28,57% and S decreased with 2,96%. S wave was significantly decreased ($p < 0,01$) whereas the E/E' ratio was significantly increased ($p < 0,001$) in the coarctation patients at rest and during exercise compared to controls. PWV, lower in coarctation patients, increased significantly with 13,99% ($p < 0,001$) during isometric exercise.
Conclusion: The results show a normal systolic and impaired diastolic LV dysfunction in children with successful early repair with no further deterioration of the diastolic function during exercise.

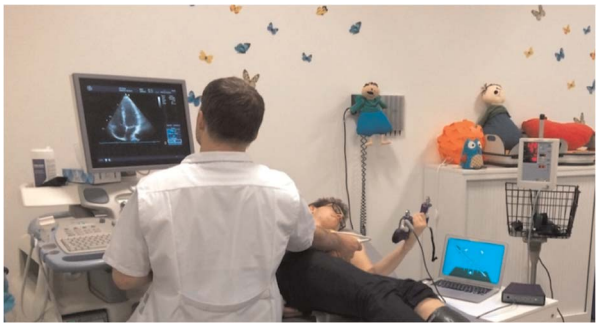

Figure 1.

P1977 - EARLY DETECTION OF MYOCARDIAL DYSFUNCTION AFTER ANTHRACYCLINE TREATMENT IN ASYMPTOMATIC PEDIATRIC CANCER PATIENTS COMPARISON OF LEFT VENTRICULAR GLOBAL STRAIN MEASURED WITH TRIPLANE AND 2D SPECKLE TRACKING

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Objective: Two-dimensional (2D) speckle tracking echocardiography (STE) are more useful to determine subclinical dysfunction. We firstly aimed to determine the left ventricular strain of pediatric cancer patients with normal ejection fraction treated with anthracycline using the 2D STE. We also compared global longitudinal strain values with using conventional 2D-STE and Triplane-STE in patients receiving anthracyclines.We also compared global longitudinal strain values with using conventional 2D-STE and Triplane-STE in patients receiving anthracyclines.
Methods: This study included 23 cross-sectionally enrolled pediatric cancer patients receiving anthracycline chemotherapy (median age: 14 years, range 6-19) and 17 controls matched for age, gender and body surface area. Patients had received a median cumulative dose of 150 mg/m2 (range 60-360 mg/m2).In all children standard 2D, M-mode, Doppler and 2D-STE and triplane-STE imaging data were obtained.
Results: 1. Patient group had a higher heart rate when compared with controls ($p = 0.016$). 2. Measurements were taken from the base of the interventricular septum; ETs (Ejection time) values were significantly decreased and MPIs values were significantly increased in patients; measurements are taken from the base of the left ventricular free wall; mS velocities were showed statistically significant difference (respectively $p = 0.022$, $p = 0.042$ and $p = 0.001$). 3. Cancer patients had a lower longitudinal and circumferential myocardial deformation of the left ventricle (respectively $p = 0.003$, $p = 0.01$). 4. Longitudinal strain values measured with Triplane method was significantly reduced in anthracycline group. Correlation between Longitudinal strain values measured with 2D and 3 P STE were also demonstrated in anthracycline group (Table 1).
Conclusion: Systolic and diastolic functions are considered in asymptomatic patients with normal ejection fraction after chemotherapy. This study confirms the subclinical LV dysfunction in patients after receiving anthracyclines with using Doppler and STE methods. We also exhibit the feasible and reproducible use of the triplane STE analysis for assessment of global LV function in the pediatric population.

Table 1. Global Longitudinal Peak Strain values of patient versus control group

GLPS	Patient group		P value	Control group		P value
(%)	2D	Triplane		2D	Triplane	
A4CH§	-18,5 ± 2,8	-18,1 ± 3,3	0,659	-20,1 ± 2,2	-19,8 ± 1,7	0,655
A3CH§	-18,6 ± 3,0	-18,1 ± 3,1	0,294	-21,4 ± 2,5	-21,6 ± 2,7	0,547
A2CH	-18,5 ± 1,8	-18,8 ± 3,5	0,904	-19,3 ± 2,3	-22,6 ± 2,4★	0,010
Avg§	-18,6 ± 2,1	-18,3 ± 2,8	0,543	-20,3 ± 1,4	-21,4 ± 1,5★	0,021

A3CH, apical 3 chamber view; A4CH, apical 4 chamber view; A2CH, apical 2 chamber view, Avg, global average GLPS; GLPS, global longitudinal peak systolic strain
Data are expressed as mean ± SD.
§$P < 0.05$ between the patient and control group ,★$P < 0.05$ between 2D and Triplane approach

P2049 - RIGHT ATRIAL MASS IN A CHILD TUMOR OR THROMBUS A CASE REPORT

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Background: One of the most common complication of indwelling central venous catheters in infants and children is right atrial thrombosis; the majority of the available research limited to case studies or retrospective small cases series. Thrombosis is associated with coexisting diseases like malignancy. In this report, it is described the presentation, treatment, technique of complete resection, and outcome of 11 years old with a thrombus vs tumor in right atrium.
Case Report: An 11 years-old boy with a diagnosis of synovial sarcoma of the pelvis treated with radiotherapy, surgical resection and chemotherapy with complete remission on computerized tomography scan. Patient had a porta-cath that had not been used or flushed for several months.Later he presented with a two weeks history with cough, chest pain on the side of the porta-catheter, shortness of breathless with minimal activity, orthopnea, exercising intolerance and limited physical activity. Computerized tomography scan showed large filling defect noted involving the right atrium crossing the tricuspid valve to the right ventricle,

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otherwise pelvic collection was solved. Transthoracic echocardiography showed a large mass (24 × 42mm) in the right atrium obstructing the right ventricle inflow with mildly depressed ventricular function. Diagnosis of right atrial mass (thrombus versus tumor) with a probably thromboembolic event was made. Patient was successfully managed with mass surgical resection. Histopathology confirmed the diagnosis of thrombus without cytology atypical or evidence of malignancy.
Conclusion: There are no pediatrics studies addressing the risk of pulmonary embolism or mortality associated with right atrial thrombus.The incidence of Right atrial thrombus in children with cancer and indwelling catheters is about 8.3% (1). Echocardiography is the primary modality for initial evaluation of symptomatic patients but definitive diagnosis requires histologic confirmation. 1.-Korones DN, Buzzard CJ, Asselin BL, Harris JP. Right atrial thrombi in children with cancer and indwelling catheters. J Pediatr .1996; 128:841-846.

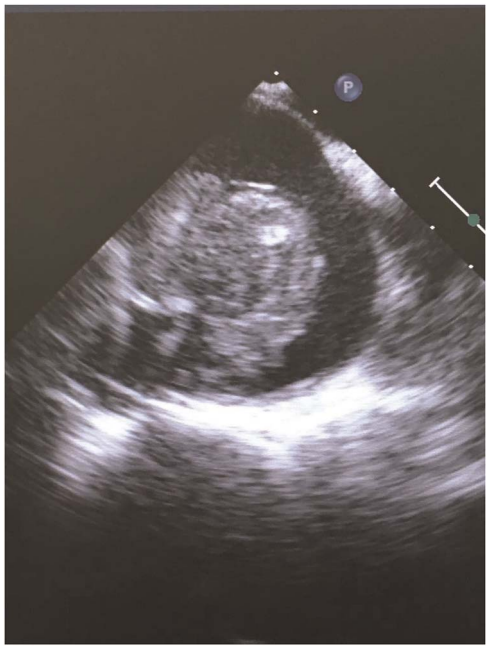

Figure.

P2050 - PRELIMINARY EVALUATION OF SYSTOLIC RV FUNCTION USING STRAIN IMAGING IN POST TOF CORRECTION PRESERVED VS NON PRESERVED PULMONARY VALVE

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National Heart Institute, Noninvasive Cardiolab, Kuala Lumpur-Malaysia[1]; National Heart Institute, Paediatric Cardiology, Kuala Lumpur-Malaysia[2]

Introduction: Right ventricle dysfunction is well known to cause significant late morbidity and mortality in post TOF correction patients and strain imaging has been an emerging tool in assessing RV function.Our aim in this preliminary study is to quantitatively evaluate systolic RV function between preserve and non-preserve pulmonary valve.
Method: 12 asymptomatic patients(9 non preserve,3 preserve pulmonary valve)were selected and underwent strain imaging using VVI. Mean age was 14.75 and median age was 15. Systolic RV strain,strain rate (SR) and longitudinal displacement were measured in 4 RV segments free wall and septum excluding the apex.
Results: Results showed no statistically significant difference between preserve and non-preserve PV in strain (-12.91% + /- 4.12 vs -14.80% + /-3.00) p = 0.48, SR (-1.08 s/1 + /- 0.46 vs -0.89 s/1 + /- 0.16) p = 0.51 and longitudinal displacement (5.30mm + /- 1.31 vs 5.96mm + /- 1.15) p = 0.45.
Conclusion: Due to limited sample size, we were not able to make a concrete conclusion based on the results between these 2 group of patients. Therefore, further investigation with larger sample size is crucial and inter relation strain imaging between right and left ventricle is recommended to further ascertain this.

P2070 - EFFICACY OF TISSUE MITRAL ANNULAR DISPLACEMENT DURING POSTOPERATIVE PERIOD IN PEDIATRIC CARDIAC SURGERY

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Background: Cardiac function temporarily decreases after heart surgery. Although echocardiography is a useful and repeatable modality, its postoperative use is limited because of the poor echo window. Tissue mitral annular displacement (TMAD) shows significant correlation with the left ventricular (LV) ejection fraction obtained by cardiac magnetic resonance imaging. We evaluated the detection rate of the available echocardiographic techniques just after operation in our pediatric cardiac surgery and compared TMAD and other index.
Methods: In this retrospective study, 111 consecutive patients with biventricular circulation were assessed using either the iE33 S8-1 or S5-1 probe within 1 h after the operation. We evaluated which index among the following was testable: TMAD, modified Simpson (four chamber view) (mS-4CV), modified Simpson (biplane) (mS-BP), and Pombo (LV short axis).
Results: Among all patients evaluated, 53 patients (48%) could be tested with TMAD, 45 (41%) with mS-4CV, nine (8%) with mS-BP, and 40 (36%) with Pombo. In median sternotomy (N = 104), the detection rate of TMAD (46%) was the highest; however, in left side thoracotomy (N = 7), the detection rate of Pombo (86%) was the highest. Finally, for 68 cases (61%), either TMAD or Pombo was possible.
Discussion: TMAD and mS-4CV were assessed by the same image; however, TMAD is useful in postoperative period. While TMAD was calculated using three tissue points, mS-4CV required all LV walls in the four chamber view. The detection rate of Pombo was low; however, combining TMAD and Pombo improved the detection rate to 61%. It resulted in the difference of based image between the four chamber and LV short axis views. Our study shows that TMAD and Pombo were useful for both the pre- and the post-operative period and provided a good evaluation rate.

P2085 - RIGHT VENTRICULAR LONGITUDINAL STRAIN FREE WALL VERSUS 4 CHAMBER STRAIN ANALYSIS. A COMPARISON IN DIFFERENT CONDITIONS

Guillermo Larios[1], Isha Crossley[2], Mieke Driessen[3], Luc Mertens[1], Andreea Dragulescu[1], Mark Friedberg[1]
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Netherlands[2]; University Medical Center Utrecht, Utrecht-The Netherlands[3]

Background: Right ventricular(RV) myocardial deformation measured by 2-D speckle tracking is useful to assess RV systolic function. There is however, no consensus on how to measure RV longitudinal strain with respect to inclusion or exclusion of the septal wall(RV-SWS). The objective of this study was to compare RV strain values analyzing only the free wall(RV-FWS) versus the RV-4-chamber (RV-4C = RV-FWS + RV-SWS) in patients with different diseases affecting the RV, in relation to RV ejection fraction(RV-EF).
Methods: We analyzed 163 patients (90 children and 73 adults) between 4-74 years with the following diagnoses: repaired Tetralogy of Fallot (TOF,n = 38), atrial septal defect (ASD,n = 14), pulmonary hypertension (PAH,n = 22), systemic RV (SysRV,n = 15) and 74 healthy controls. Two observers traced the RV 6 segments using a RV-centered 4-chamber view. The peak(PK) and end-systolic(ES) strain were recorded. We calculated the difference between RV-FWS(3-segments) and RV-4C(6-segments) and correlated them with cardiac magnetic resonance derived RV-EF.
Results: RV-FWS, RV-SWS and RV-4C for each group are presented in the Table. The highest RV-FWS and RV-4C was seen in ASD and the lowest in the SysRV group. The largest difference between RV-FW and RV-4C analysis was in the ASD group and the smallest in the ToF group. Small, but significant differences were found between PK and ES strain (paired t-test in RV-4C, $p < 0.05$ in all groups except for SysRV). The best correlation with RV-EF by group using RV-FW/RV-4C were: Controls: RV-4C(ES), r = -0.28, p = 0.06, ToF: RV-4C(ES), r = -0.45, p = 0.01, ASD: no correlation, PAH: RV-FWS(PK), r = -0.73, p = 0.01 and SysRV: RV-FWS(ES), r = -0.8, $p < 0.01$.
Conclusions: The RV-FWS and RV-4C yield different results and correlation with RV-EF depending on the underlying disease. This may reflect the different timing and magnitude of septal function in different conditions. In patients exposed to high RV pressures, RV-EF correlates more closely with RV-FWS than with RV-4C. Uniform approach to RV strain analysis may not yield optimal data across all conditions.

Table 1. Demographic, conventional echocardiographic parameters, CMR and 2D speckle tracking strain analysis according to RV condition. Data presented as mean ± standard deviation

	Controls	TOF	ASD
N	74	38	14
Gender: Male, %	39 (52.7%)	24 (63.2%)	9 (64.3%)
Age, years	21.5 ± 12.6	19.4 ± 9.4	10.2 ± 3.9
Height, cm	167.4 ± 20.5	163.5 ± 18.3	142.2 ± 23.4
Weight, kg	61 ± 18.7	61.2 ± 19.3	38.4 ± 17.3
Body Surface Area, m^2	1.67 ± 0.36	1.66 ± 0.36	1.21 ± 0.38
Conventional echocardiogram RV functional parameters			
TAPSE, mm	21.9 ± 3.2	15.3 ± 4	27.4 ± 3.8
TDI S' RV lateral wall, cm/s	14.7 ± 2.7	9.8 ± 2.4	17.7 ± 2.4
RV end diastolic area, cm^2	22.8 ± 5.7	39.4 ± 9.6	31 ± 6
RV FAC, %	48.4 ± 5.5	37.6 ± 8.3	50.3 ± 4.6
CMR			
RV end diastolic volume index, ml/m^2	106.2 ± 15.1	156.2 ± 40.1	157.5 ± 41*
RV end systolic volume index, ml/m^2	53.3 ± 8.8	91.8 ± 31.9	67.8 ± 13.9*
RV ejection fraction, %	50.4 ± 3.9	41.8 ± 7.5	56.2 ± 4.2*
2D-speckle tracking strain			
RV-SWS average (peak), %	-17 ± 2.9	-15.9 ± 4.4	-21.9 ± 4.2
RV-SWS average (end syst), %	-16.9 ± 2.9	-15.9 ± 4.3	-21.8 ± 4.3
RV-FWS average (peak), %	-25.7 ± 4.1	-17.5 ± 7.5	-31.1 ± 4.5
RV-FWS average (end syst), %	-25.5 ± 4.1	-17.7 ± 4.7	-30.2 ± 4.3
RV-4C average (peak), %	-21.3 ± 3.1	-17.2 ± 4.3	-26.4 ± 3.9
RV-4C average (end systole), %	-21.2 ± 3	-16.9 ± 4.2	-26 ± 3.9
FWS - 4C (peak), %	-4.4 ± 1.8	-0.3 ± 5.7	-4.7 ± 2.1
FWS - 4C (end systole), %	-4.3 ± 1.8	-0.8 ± 1.7	-4.2 ± 1.9
RV-FWSl strain rate (peak), s^{-1}	-1.5 ± 0.5	-1 ± 0.4	-2 ± 0.5
RV-4C strain rate (peak), s^{-1}	-1.6 ± 0.3	-1.2 ± 0.3	-1.8 ± 0.4

Table 1. *Continued*

	PAH	Systemic RV	Total
N	22	15	163
Gender: Male, %	13 (59.1%)	10 (66.7%)	95 (58.3%)
Age, years	33.4 ± 26.3	35.7 ± 7.5	22.9 ± 15.3
Height, cm	148.3 ± 31.9	175.9 ± 7.8	162.4 ± 23.2
Weight, kg	53.3 ± 28.8	74.7 ± 7.1	59.3 ± 21.2
Body Surface Area, m^2	1.42 ± 0.56	1.91 ± 0.12	1.62 ± 0.41
Conventional echocardiogram RV functional parameters			
TAPSE, mm	14.8 ± 3.3	-	19.6 ± 6.3
TDI S' RV lateral wall, cm/s	9.4 ± 2.4	-	12.7 ± 4.2
RV end diastolic area, cm^2	-	-	32.4 ± 10.3
RV FAC, %	30.5 ± 11.3	-	41.9 ± 10.5
CMR			
RV end diastolic volume index, ml/m^2	149.2 ± 33.1	136.3 ± 45.3	136.2 ± 40.3
RV end systolic volume index, ml/m^2	104.8 ± 31.1	89.2 ± 42.6	76.8 ± 32.1
RV ejection fraction, %	31.9 ± 8	36.8 ± 8.7	44.9 ± 9.6
2D-speckle tracking strain			
RV-SWS average (peak), %	-12 ± 4.4	-10.6 ± 2.6	-15.9 ± 4.6
RV-SWS average (end syst), %	-12 ± 4.4	-10.6 ± 2.6	-15.8 ± 4.6
RV-FWS average (peak), %	-18.1 ± 4.6	-14.1 ± 4.3	-22.2 ± 7.2
RV-FWS average (end syst), %	-17.6 ± 4.5	-14 ± 4.6	-22 ± 6.6
RV-4C average (peak), %	-15.2 ± 4	-12.4 ± 3.4	-19.2 ± 5.2
RV-4C average (end systole), %	-14.9 ± 4	-12.4 ± 3.5	-18.9 ± 5.1
FWS - 4C (peak), %	-2.9 ± 1.7	-1.7 ± 1.8	-3 ± 3.6
FWS - 4C (end systole), %	-2.6 ± 1.8	-1.6 ± 1.8	-3 ± 2.3
RV-FWSl strain rate (peak), s^{-1}	-1.1 ± 0.5	-0.6 ± 0.2	-1.3 ± 0.6
RV-4C strain rate (peak), s^{-1}	-1.3 ± 0.5	-	-1.5 ± 0.4

Abbreviations (not mentioned in the abstract): TDI: Tissue Doppler imaging; TAPSE: Tricuspid annular plane systolic excursion. *: Right ventricle volumes estimation using 2-D echocardiogram based 3-D reconstruction method (Ventripoint®, Seattle, USA)

Table 2. Demographic, conventional echocardiography parameters, CMR and 2D speckle tracking strain analysis according to RV condition. Data presented as mean ± standard deviation

	Controls	TOF	ASD	PAH	Systemic RV	Total
N	74	38	14	22	15	163
Gender: Male, %	39 (52.7%)	24 (63.2%)	9 (64.3%)	13 (59.1%)	10 (66.7%)	95 (58.3%)
Age, years	21.5 ± 12.6	19.4 ± 9.4	10.2 ± 3.9	33.4 ± 26.3	35.7 ± 7.5	22.9 ± 15.3
Height, cm	167.4 ± 20.5	163.5 ± 18.3	142.2 ± 23.4	148.3 ± 31.9	175.9 ± 7.8	162.4 ± 23.2
Weight, kg	61 ± 18.7	61.2 ± 19.3	38.4 ± 17.3	53.3 ± 28.8	74.7 ± 7.1	59.3 ± 21.2
Body Surface Area, m^2	1.67 ± 0.36	1.66 ± 0.36	1.21 ± 0.38	1.42 ± 0.56	1.91 ± 0.12	1.62 ± 0.41
Conventional echocardiogram RV functional parameters						
TAPSE, mm	21.9 ± 3.2	15.3 ± 4	27.4 ± 3.8	14.8 ± 3.3	-	19.6 ± 6.3
TDI S' RV lateral wall, cm/s	14.7 ± 2.7	9.8 ± 2.4	17.7 ± 2.4	9.4 ± 2.4	-	12.7 ± 4.2
RV end diastolic area, cm^2	22.8 ± 5.7	39.4 ± 9.6	31 ± 6	-	-	32.4 ± 10.3
RV FAC, %	48.4 ± 5.5	37.6 ± 8.3	50.3 ± 4.6	30.5 ± 11.3	-	41.9 ± 10.5
CMR						
RV end diastolic volume index, ml/m^2	106.2 ± 15.1	156.2 ± 40.1	157.5 ± 41*	149.2 ± 33.1	136.3 ± 45.3	136.2 ± 40.3
RV end systolic volume index, ml/m^2	53.3 ± 8.8	91.8 ± 31.9	67.8 ± 13.9*	104.8 ± 31.1	89.2 ± 42.6	76.8 ± 32.1
RV ejection fraction, %	50.4 ± 3.9	41.8 ± 7.5	56.2 ± 4.2*	31.9 ± 8	36.8 ± 8.7	44.9 ± 9.6
2D-speckle tracking strain						
RV-SWS average (peak), %	-17 ± 2.9	-15.9 ± 4.4	-21.9 ± 4.2	-12 ± 4.4	-10.6 ± 2.6	-15.9 ± 4.6
RV-SWS average (end syst), %	-16.9 ± 2.9	-15.9 ± 4.3	-21.8 ± 4.3	-12 ± 4.4	-10.6 ± 2.6	-15.8 ± 4.6
RV-FWS average (peak), %	-25.7 ± 4.1	-17.5 ± 7.5	-31.1 ± 4.5	-18.1 ± 4.6	-14.1 ± 4.3	-22.2 ± 7.2
RV-FWS average (end syst), %	-25.5 ± 4.1	-17.7 ± 4.7	-30.2 ± 4.3	-17.6 ± 4.5	-14 ± 4.6	-22 ± 6.6
RV-4C average (peak), %	-21.3 ± 3.1	-17.2 ± 4.3	-26.4 ± 3.9	-15.2 ± 4	-12.4 ± 3.4	-19.2 ± 5.2
RV-4C average (end systole), %	-21.2 ± 3	-16.9 ± 4.2	-26 ± 3.9	-14.9 ± 4	-12.4 ± 3.5	-18.9 ± 5.1
FWS - 4C (peak), %	-4.4 ± 1.8	-0.3 ± 5.7	-4.7 ± 2.1	-2.9 ± 1.7	-1.7 ± 1.8	-3 ± 3.6
FWS - 4C (end systole), %	-4.3 ± 1.8	-0.8 ± 1.7	-4.2 ± 1.9	-2.6 ± 1.8	-1.6 ± 1.8	-3 ± 2.3
RV-FWSl strain rate (peak), s^{-1}	-1.5 ± 0.5	-1 ± 0.4	-2 ± 0.5	-1.1 ± 0.5	-0.6 ± 0.2	-1.3 ± 0.6
RV-4C strain rate (peak), s^{-1}	-1.6 ± 0.3	-1.2 ± 0.3	-1.8 ± 0.4	-1.3 ± 0.5	-	-1.5 ± 0.4

Abbreviations (not mentioned in the abstract): TDI: Tissue Doppler imaging; TAPSE: Tricuspid annular plane systolic excursion. *: Right ventricle volumes estimation using 2-D echocardiogram based 3-D reconstruction method (Ventripoint®, Seattle, USA)

P2090 - ETHICAL CONSIDERATIONS IN FETAL DIAGNOSIS OF CONGENITAL HEART DISEASE A NARRATIVE REVIEW

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Background/Hypothesis: Prenatal examination with fetal echocardiography has increasingly become the primary mode of diagnosis for most congenital heart disease. This shift in diagnosis from postnatal to prenatal, in turn, has altered conversations regarding management discussions, prognostication, and treatment options to prior to birth. Given that these discussions can be based on incomplete information and evolving disease processes, ethical dilemmas can and often do arise. This narrative review aims to evaluate the existing evidence on ethical dilemmas encountered in the prenatal diagnosis of congenital heart disease.
Methods: We searched MEDLINE via PubMed, Embase, and Google Scholar to identify studies assessing ethical issues in fetal diagnosis of congenital heart disease. We used controlled vocabulary when possible; search terms included: "ethics," "ethical," "fetus," "fetal," "cardiology," "prenatal counseling", "echocardiography," and "congenital heart disease."
Results: We identified 61 articles spanning the years 1983–2016. Fourteen studies were published between 1983–1999, 22 between 2000–2009, and 25 between 2010–2016. We identified 4 categories of articles: 23 opinion/commentary/ethical analysis, 16 review papers, 19 cross-sectional survey studies, and 3 case-series papers. On analysis of the studies, the following ethical themes emerged: what comprises appropriate counseling for families, the factors that influence counseling and decision making including termination of pregnancy, fetal cardiac intervention, and the influence of concomitant genetic diagnosis in decision making.
Conclusion: There is a critical need to explore ethical questions related to the fetal diagnosis of congenital heart disease. There is currently a paucity of data in this area. Existing literature is mainly comprised of opinion, commentary, and review papers. Future studies should focus on evidence-based approaches to understanding the emerging ethical issues identified in this narrative review.

P2091 - CHARACTERISTICS AND OUTCOMES IN A SPANISH REGISTRY FOR CHILDREN WITH BICUSPID AORTIC VALVE (REVAB)

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Objectives: Define demographics, valve morphology, associated diseases and complications in a nationwide registry.
Methods: Online registry under the auspices of Spanish Society of Pediatric Cardiology and Congenital Heart Diseases (SECPCC) started on May 2016 for pediatric patients (<18 years of age) with bicuspid aortic valve.
Results: A total of 944 patients from 28 hospitals were introduced by November 2016. Mean age at diagnosis was 3.0 ± 4.0years, 52.3% before 1 year of age. There was a prenatal suspicion of congenital heart disease in 9.9% of patients. Family history of bicuspid aortic valve was known for 6.3% of patients. The majority were male, 68.6%. Valve morphology was horizontal in 52.5% and only 34.1% were pure. A total of 589 (62.4%) had an isolated bicuspid aortic valve, 265 (28.1%) had concomitant left sided obstructive lesions (including coarctation of the aorta, interrupted aortic arch, subaortic stenosis or mitral stenosis), 121 ventricular septal defect (12.8%); and others (55, 5.8%). A total of 281 patients (29.7%) had 380 interventions: 263 on the aortic arch (216 surgical, 47 percutaneous), 94 aortic valvuloplasty (77 percutaneous, 17 surgical), 10 mechanical aortic valves, 8 Ross, 2 Bentall, 1 Yacoub and 2 substitution of the ascending aorta. At a mean age of 7.2 ± 4.9years, 166 patients (17.6%) had $\geq$ moderate aortic stenosis and 62 (6.5%) had $\geq$ moderate aortic regurgitation. The mean aortic root and the ascending aorta were 0.1 ± 1.7 SD and 1.1 ± 2.2 SD respectively, 8% had dilatation of the aortic root and 25.8% of the ascending aorta (z-score >2). Mean progression of the aortic root and the ascending aorta was 0.2 ± 0.6 SD/y and 0.3 ± 0.7 SD/y respectively. A total of 20 endocarditis have been reported (2.1%), 1 aortic dissection and 9 exitus (median 2 months, range birth to 13.8years).
Conclusions: Pediatric patients with bicuspid aortic valve are not exempt from complications during childhood, thus close follow-up is warranted.

P2096 - EARLY CHANGES IN LEFT VENTRICLE MYOCARDIAL FUNCTION AND MASS IN OBESE NON–HYPERTENSIVE MEXICAN CHILDREN

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Background: Previous studies have reported structural and functional changes of left ventricle (LV) associated with adult obesity, and abnormalities of LV longitudinal myocardial deformity in severely obese adolescents. The aim of this study is to evaluate early changes in LV myocardial function in children with obesity without hypertension.
Methods: We performed a cross-sectional study in children between 6-15 years old, who underwent anthropometric, biochemical and LV function evaluation by conventional bi and three-dimensional echocardiography, and analysis of longitudinal myocardial deformity using the Speckle Tracking technique. The comparison between groups was performed using the ANOVA test, and multiple comparisons with the Bonferroni method.
Results: 133 children were included, with an average age of 9.5 years. They were divided into three groups according to their body mass index Z score: normal weight (n = 49), overweight

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(n = 33), and obesity (n = 51). The obesity group showed lower blood levels of HDL cholesterol (45 mg/dl vs. 58 mg/dl, $p < 0.05$) and higher levels of triglycerides (130 mg/dl vs. 76 mg/dl, $p < 0.05$) compared to the normal weight group. The obesity group showed a higher left atrium diameter (28.4mm vs. 25.7mm, $p < 0.05$), LV diastolic diameter (41.9mm vs. 25.7mm, $p < 0.05$), LV 2Dmass (103.4 g vs. 75.6 g, $p < 0.05$), and LV 3Dmass (86.7 g vs. 60.4 g, $p < 0.05$) in comparison with the normal weight group. Conventional indicators of LV function, including ejection fraction, showed no significant differences between groups, while the global myocardial deformation was lower among obese children than normal weight group (-20.0% vs. -23.8%, $p < 0.05$). There were no significant differences between overweight and normal weight group. *Conclusions:* These results support the relevance of echocardiographic methods to assess the function, anatomic structure and myocardial deformation of the left ventriclein pediatric patients with obesity. Although overweight was not associated with significant changes, it is important to avoid its progress towards obesity to prevent early cardiovascular abnormalities.

Table 1. General characteristics of all three groups of study

	Normal Weight (n = 49)	Overweight (n = 33)	Obesity (n = 51)	P value
Age (years)	9.4 ± 2.5	9.4 ± 2.0	9.8 ± 1.8	0.688
BMI Z score (Kg/(m)^2)	0.17 ± 1.1	1.44 ±0.3 $^{\alpha}$	2.81 ±0.6 $^{\beta}$	<0.001
SBP (mmHg)	94 ± 10.8	97 ± 8.1	97 ± 21.5	0.672
DBP (mmHg)	62 ± 8.3	65 ± 5.5	62 ± 16.7	0.480
Glu (mg/dl)	90 ± 6.3	90 ± 4.2	91 ± 5.4	0.556
TC (mg/dl)	159 ± 24.2	163 ± 26.9	164 ± 26.2	0.669
HDL (mg/dl)	58 ± 12.8	50 ± 12.2$^{\alpha}$	45 ± 9.1$^{\beta}$	<0.001
LDL (mg/dl)	95 ± 22.2	103 ±24.7	128 ± 149.4	0.211
TGL (mg/dl)	76 ± 34.4	92 ± 38.8	130 ± 41.8$^{\beta}$	<0.001

BMI: body mass index. SBP: Systolic blood pressure. DBP: Diastolic blood pressure. Glu: glucose. TC: total cholesterol. LDL: Low-density cholesterol. HDL: High-density cholesterol. TGL: triglycerides. The comparison between groups was performed using the ANOVA test and multiple comparisons with the Bonferroni method, were **β** $p < 0.05$ was used for the comparison between normal weight versus obesity group, and **α** $p < 0.05$ for normal weight versus overweight group.

Table 2. Left ventricular structure dimensions and mass of each group.

	Normal Weight (n = 49)	Overweight (n = 33)	Obesity (n = 51)	P value
LA (mm)	25.7 ± 3.1	26.9 ± 4.6	28.4 ± 4.8$^{\beta}$	0.013
LA Z score	0.96 ± 0.86	0.89 ± 1.04	0.83 ± 1.08	0.821
LVDD (mm)	38.8 ± 3.6	41.8 ± 4.5$^{\alpha}$	41.9 ± 4.4$^{\beta}$	0.001
LVDD Z score	-0.5 ± 0.68	-0.19 ± 0.79	-0.56 ± 0.95	0.109
LVSD (mm)	24.6 ± 2.7	25.9 ± 4	26.3 ± 5.2$^{\beta}$	0.141
LVSD Z score	-0.2 ± 0.85	-0.31 ± 0.84	-0.2 ± 0.94	0.841
IVSD (mm)	6.7 ± 1.2	7.1 ± 1.5	7.4 ± 1.6$^{\beta}$	0.061
SIVD Z score	0.29 ± 0.78	0.47 ± 0.74	0.16 ± 1.5	0.523
LVPW (mm)	5.9 ± 1.3	6.6 ± 1.1	6.6 ± 1.9$^{\beta}$	0.046
LVPW Z score	0.38 ± 0.87	0.33 ± 1	0.13 ± 1	0.433
LV Mass (g)	75.6 ± 26.7	81.7 ± 26.1	103.4 ± 29.9$^{\beta}$	< 0.001
LV Mass Z score	-0.79 ± 1.1	-0.56 ± 1.15	0.3 ± 1.1$^{\beta}$	< 0.001
LV Mass (g/m^2)	67 ± 21.3	66 ± 13.8	71.1 ± 13.3	0.357

LA: left atrium. LVDD: left ventricular diastolic diameter. LVSD: left ventricular systolic diameter. IVSD: interventricular septum diastolic diameter. LVPW: left ventricular posterior wall. LV: left ventricle. The comparison between groups was performed using the ANOVA test and multiple comparisons with the Bonferroni method, were **β** $p < 0.05$ was used for the comparison between normal weight versus obesity group, and **α** $p < 0.05$ for normal weight versus overweight group.

Table 3. Left ventricular parameters of systolic and diastolic function of each group.

	Normal weight (n = 41)	Overweight (n = 19)	Obesity (n = 41)	P value
LVEF (%)	64.6 ± 4.7	64.7 ± 5.3	64.1 ± 6	0.855
LVSF (%)	35.8 ± 5.2	38.7 ± 7.5	38.2 ± 9	0.168
Em/Am Ratio	1.7 ± 0.31	2.2 ± 1.9	1.6 ± 0.32$^{\beta}$	0.043
Em/Am Ratio Z score	-0.31 ± 0.63	-0.12 ± 0.65	-0.35 ± 0.75$^{\beta}$	0.401
LVS MPI	0.39 ± 0.08	0.4 ± 0.08	0.45 ± 0.45	0.662
LVS MPI Z score	0.35 ± 0.84	2.68 ± 11.1	0.53 ± 1.19	0.176
LVM MPI	0.37 ± 0.07	0.38 ± 0.09	0.41 ± 0.11	0.235
LVM MPI. Z score	0.26 ± 0.86	0.48 ± 0.93	0.64 ± 1.27	0.262

LVEF: left ventricular ejection fraction. LVSF: left ventricular shortening fraction. Em/Am Ratio: e wave and a wave relation. LVS MPI: left ventricular septum myocardial performance index. LVM MPI: left ventricular mitral myocardial performance index.
The comparison between groups was performed using the ANOVA test and multiple comparisons with the Bonferroni method, were
β $p < 0.05$ was used for the comparison between normal weight versus obesity group, and **α** $p < 0.05$ for normal weight versus overweight group.

Table 4. Left ventricular longitudinal myocardial deformity, represented in segments and global strain, of all study groups.

Segment	Normal Weight (n = 49)	Over-weight (n = 33)	Obesity (n = 51)	P value
1	-21 ± 5.6	-24 ± 7.4	-21 ± 8.2	0.082
2	-19 ± 3	-20 ± 5.1	-19 ± 4.9	0.548
3	-19 ± 4.1	-21 ± 4.4	-18 ± 4.8	0.092
4	-21 ± 4.3	-23 ± 5.1	-18 ± 6$^{\beta}$	0.001
5	-24 ± 4.7	-24 ± 5.5	-22 ± 3.8	0.327
6	-22 ± 3.6	-24 ± 5.1	-21 ± 8.7	0.075
7	-23 ± 6	-23 ± 6.6	-19 ± 5.8$^{\beta}$	0.002
8	-22 ± 4.1	-25 ± 6.8	-22 ± 5.3	0.098
9	-22 ± 3.2	-23 ± 5.6	-21 ± 4.8	0.170
10	-21 ± 4.5	-22 ±4.8	-19 ± 5.2$^{\beta}$	0.015
11	-23 ± 4.2	-22 ± 4.3	-21 ± 5	0.313
12	-24 ± 4.3	-23 ± 6.3	-22 ± 6.1	0.375
13	-24 ± 5.7	-20 ± 5.7$^{\alpha}$	-18 ± 5$^{\beta}$	<0.001
14	-26 ± 4.8	-23 ± 5.3$^{\alpha}$	-23 ± 3.9$^{\beta}$	0.004
15	-26 ± 5.4	-25 ± 5.8	-23 ± 3.9$^{\beta}$	0.015
16	-24 ± 3.3	-21 ± 4.3$^{\alpha}$	-20 ± 5$^{\beta}$	<0.001
17	-25 ± 3.7	-22 ± 4.2$^{\alpha}$	-20 ± 3.4$^{\beta}$	<0.001
LGS (%)	-23.5 ± 2.2	-22.2 ± 2.6	-20 ± 2.3$^{\beta}$	<0.001

LGS: longitudinal global strain. The comparison between groups was performed using the ANOVA test and multiple comparisons with the Bonferroni method, were **β** $p < 0.05$ was used for the comparison between normal weight versus obesity group, and **α** $p < 0.05$ for normal weight versus overweight group.

P2098 - SPECTRUM OF THROMBOEMBOLIC MANIFESTATIONS IN CONGENITAL CYANOTIC HEART DISEASES

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Background: Thromboembolic manifestations are common in congenital cyanotic heart diseases (CCHD) due to high haematocrit,coagulation abnormalities, low platelet counts and an unpredictable response to fluid shifts. We present nine patients with varied manifestations.
Materials and Methods: Children with CCHD presenting with thromboembolic manifestations at Rabindranath Tagore International Institute of Cardiac Sciences and Narayana Superspeciality Hospital, during the period July 2014 – June 2016, formed the study group.
Results: The background CCHD resulting in the manifestations were Tetralogy of Fallot, Pulmonary atresia/VSD and single ventricle - pulmonary stenosis. 5 were males and 4 were females, with age range of 6 months to 16 years. The different presentations were: prolonged fever and brain abscesses (2), occipital infarct resulting in cortical blindness (1), severe pain abdomen due to splenic infarct (1), portal vein thrombosis (1), medullary infarct resulting in cranial nerve palsy and nasal regurgitation (1), post diarrhoeal irritability with complete occlusion of the superior sagittal sinus and raised intracranial tension (1), hyperpyrexia following cardiac catheterisation (1), fever and convulsions with bilateral thalamic infarcts generalising to extensive bilateral cerebral infarcts (1). Children with brain abscesses were treated with abscess drainage and antibiotics for six weeks. Children with occipital infarct, splenic infarct and medullary infarct, were treated conservatively. The children with superior sagittal sinus thrombosis and portal vein thrombosis had warfarin therapy and complete dissolution of the clots were noted in 3 months. The child with hyperpyrexia after cardiac catheterisation passed away shortly after the incident and the child with thalamic infarcts died 7 days after onset of symptoms.
Conclusions: Thromboembolic episodes present with myriad manifestations. Anticipation of atypical signs and symptoms may save the patient with appropriate therapy on time. Imaging of the concerned body part should be prompt, and repeated if necessary, to understand the lesions in details and their evolution, to guide the therapy.

P2101 - RATE AND TRENDS IN PRENATAL DETECTION OF COMPLEX CONGENITAL HEART DISEASE A SINGLE CENTER EXPERIENCE

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Background: Complex congenital heart disease (CCHD) is a major cause of morbidity and mortality in the pediatric population. Incidence of congenital cardiac anomaly is 10 per 1000 live births, 30% of which is CCHD. Fetal echocardiography is the main modality of prenatal identification of CCHD. The objective of this study is to define the rate and trends in prenatal detection of CCHD at our center.
Methods: Retrospective chart review of patients with CCHD, born in Jackson Health System, between January 2000 and November 2016, was done. Presence or absence of prenatal diagnosis, indication for fetal echocardiogram, gestational age at diagnosis, and outcome at 1-month of age were analyzed.
Results: A total of 350 CCHD patients were born during the study period, representing 3.1 per 1000 live births. The overall prenatal diagnosis rate of CCHD was 65.6%. The rate of diagnosis increased over the study period from low 50's early on to mid-high 80's near the end of the study. Lowest detection rate was noted for total anomalous pulmonary venous return (25%), coarctation of the aorta (44%), tetralogy of Fallot (48%) and transposition of great arteries (54%). Highest detection rate was noted for tricuspid atresia (90%) and unbalanced AV septal defect (83%). The most common indication for a fetal echocardiogram was abnormal prenatal ultrasound (78.8%). There was no mortality benefit at 1 month associated with prenatal diagnosis.
Conclusions: There is increasing rate of prenatal diagnosis of CCHD over the study period. This can be attributed to changes in the protocol of obstetric screening ultrasound and indications for fetal echocardiogram. In spite of these improving detection rates, certain CCHD have a lower rate of detection. Though there is no significant advantage in 1-month mortality in the prenatal detection group, there might be significant difference in early and late morbidity.

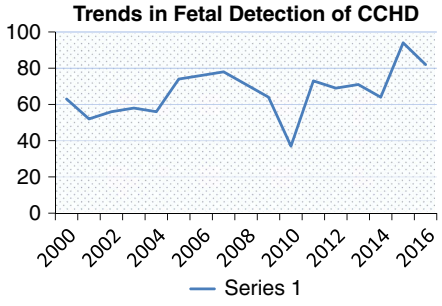

Figure 1.

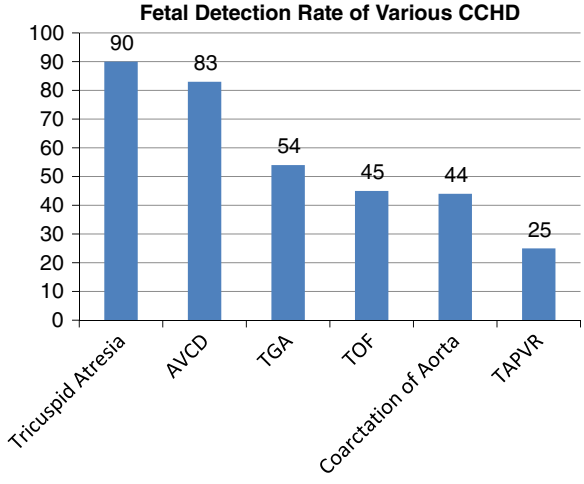

Figure 2.

P2112 - IN UTERO PROGNOSTICATING FETAL ECHOCARDIOGRAPHIC MARKERS IN ABSENT PULMONARY VALVE SYNDROME A CASE SERIES

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Background: Tetralogy of Fallot with Absent Pulmonary Valve is a rare but a well-described complex cardiac lesion that can be reliably diagnosed on prenatal ultrasound. Risk stratification and the identification of infants requiring perinatal resuscitation, however remains to be defined.
Methodology: The fetal echocardiography database of a KK Women's and Children's Hospital, a tertiary center was reviewed retrospectively over a 5 year period from 2011 to 2016 and cases of Tetralogy of Fallot with Absent Pulmonary Valve were identified, the echocardiographic data and clinical histories were analyzed.
Results: During the study period, 5 cases of Tetralogy with Absent Pulmonary Valve were diagnosed prenatally. There was severe pulmonary regurgitation with normal biventricular function in all patients. The pulmonary regurgitation VTI ranged from 16.2 to 45.9 cm. The main pulmonary artery to aortic annulus ratio ranged from 1.12–1.50. 2 patients were noted have arterial ducts. There were 2 pregnancy terminations and 1 neonatal death. None of the fetuses developed hydrops. All 3 patients who had been followed-up in utero required intubation and ventilation at birth. The infant who died in the neonatal period had an associated cystadenomatoid malformation of the left lung. 1 patient had DiGeorge Syndrome.
Conclusion: Tetralogy of Fallot with Absent Pulmonary Valve remains a critical congenital heart disease with all live-born infants requiring resuscitation at birth in our series. The fetuses with associated extra cardiac conditions tended to have a worse prognosis. The ratio of main pulmonary arterial dimension to aortic annulus dimension measured in systole was greater in neonates requiring respiratory support. Indexed measurements and Doppler derived indices during fetal life may help in further risk stratification.

P2114 - PERICARDIAL CYST IN A 3 YEAR OLD BOY A DIAGNOSTIC AND THERAPEUTIC CHALLENGE

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The authors present a 3 year old boy, who underwent a chest X-ray for the first time due to respiratory infection. The chest X-ray revealed an oval shaped mass along the left contour of the heart. It was thought to be a pneumonia and was treated with antibiotics. The following year, the boy had influenza and the chest X-ray was done again. Again it showed the same formation along the left contour of the heart above the apex. Various diagnostic procedures were done to determine pathology. The microbiological samples did not prove TBC or any other infection, a bronchoscopy excluded bronchial anomalies or a foreign body in the bronchi. An echocardiography has been made and the heart was found to be morphologically and functionally normal, but the cyst was seen along the left ventricle sized 9 × 19 mm and filled with fluid, without compression effects on the surrounding structures. A cardiac MRI confirmed the diagnosis of pericardial cyst in left lung lingui. Since the boy was asymptomatic and the control cardiac MRI and the echocardiography showed milder regression in size - to now we have not decided to treat the patient surgically. The pericardial cyst is rare, occurring with a frequency of 1 in 100 000 people and is usually an incidental finding. It makes 7% of all the masses in the mediastinum and 33% of mediastinal cysts. In early childhood it is even more rare, and may represent a diagnostic challenge in distinguishing the pericardial cyst from other, less benign mediastinal masses. For smaller cysts and asymptomatic patients most authors recommend only follow up by series of echocardiography and cardiac MRI as we did in our patient. Our boy is doing well, still is asymptomatic after 5 years follow up

P2124 - CARDIAC FUNCTION IN PEDIATRIC PATIENTS WITH CONGENITAL ADRENAL HYPERPLASIA

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Background: Congenital adrenal hyperplasia (CAH) is characterized by cortisol deficiency and hyperandrogenism. Both hyperandrogenism and treatment with supraphysiological doses of glucocorticoids may cause unfavorable changes in the cardiovascular risk profile of CAH patients. Data on the cardiac function in CAH patients are scarce.
Methods: The cardiac function of 27 pediatric CAH patients, aged 8-16 years, was evaluated by conventional echocardiography and 2D myocardial strain (rate) imaging. Results were compared to a selected cohort of 27 age and gender matched healthy controls. Data on hydrocortisone dosage in CAH patients were collected. Blood was withdrawn in CAH patients to evaluate 17-OH-progesterone and androstenedione concentrations.
Results: Left ventricular parameters (IVSd, LVIDd, LVIDs, EF and FS) are normal in all CAH patients, and do not differ from controls. In contrast, LVPWd is significantly lower in CAH patients compared to controls (5.55 vs 6.53 mm; P = 0.009). The LVPWd Z-score is significantly lower in CAH patients yet within the normal range (-1.12 vs -0.35; P = 0.002). LV mass and LV mass index are normal and comparable in both patients and controls. Isovolumetric relaxation time is significantly lower in CAH patients (49 vs 62 ms; P = 0.003). No associations were found between left ventricular parameters and BMI SDS, hydrocortisone dose, androstenedione and 17-OH-progesterone, respectively. Global longitudinal, radial and circumferential strain were normal compared to controls. Global radial strain rate was significantly higher compared to healthy controls (2.58 vs 2.06 1/s). Time to peak global longitudinal, radial and cicumferential strain did not differ between CAH patients and controls.
Conclusion: Cardiac evaluation of pediatric CAH patients showed no signs of left ventricular hypertrophy or ventricular dilatation. We found a thinner LVPWd in CAH patients, this finding was not associated with treatment or hyperandrogenism. A shorter isovolumetric relaxation time in CAH patients suggested increased left atrial pressure.

P2159 - DOPPLER ANALYSIS OF FETAL PULMONARY VENOUS FLOW IN HYPOPLASTIC LEFT HEART SYNDROME OUR EXPERIENCE IN THE LAST FIVE YEARS

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Background: Hypoplastic left heart syndrome (HLHS) is a rare heart defect that results in a significant increase in morbidity and mortality. Advances in fetal diagnosis, as well as changes in the legislation regarding the interruption of pregnancy, have reduced the incidence of this entity in newborns. Several studies suggest that the fetal pulmonary venous flow pattern relates to the severity and outcome of these patients.

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Materials and Methods: We did a retrospective review of prenatal diagnosis of HLHS in the last 5 years. An analysis of fetal pulmonary venous doppler was carried out. According to the literature, three patterns were defined: Type A, continuous forward flow; Type B, reversal A-wave; and Type C, bidirectional flow.
Results: 21 prenatal diagnosis of HLHS were reviewed. In 14 cases pregnancy was terminated by parental choice (66%). In the 7 cases remaining, fetal pulmonary venous flow pattern was analyzed: 3 A, 3B and 1 C. Delivery at term in all cases. None of the newborns required reanimation at birth. All of them were admitted to neonatal ICU. In one patient (A type) therapeutic effort was limited because of parental decision. The six remaining patients were surgically intervened (Norwood-Sano) between the 5-7th days of life. Only two infants (1B, 1 C) required balloon atrial septostomy before surgery. Both required ECMO after intervention and died a few days later due to refractory hypoxemia. The four other patients reached the surgery without any complications.
Conclusions: - HLHS is an entity with a high rate of pregnancy interruption, this limits the appearance of the most severe clinical presentations. - Abnormal fetal pulmonary venous flow (especially Type C) may be associated with refractory hypoxemia after surgery and a poorer outcome. - The doppler analysis of fetal pulmonary veins should be performed in all cases of HLHS, in order to improve prognostic information to the parents

P2164 - THE ROLE OF INTRAOPERATIVE EPICARDIAL ECHOCARDIOGRAPHY IN PEDIATRIC CARDIAC SURGERY

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Background: Recently the need for intraoperative echocardiography (transesophageal or epicardial) has been increased especially during congenital cardiac surgeries. In case of improper conditions for transesophageal echocardiography (TEE) like low body weight, inappropriate anatomy of the jaw, large tongue etc. intraoperative epicardial echocardiography (IEE) can be performed. We aimed to evaluate the findings and results of IEE patients.
Materials and Methods: Patients who had IEE between December 2015-December 2016 were analysed retrospectively. The age, gender, body weight, preoperative transthoracic echocardiography findings and IEE reports were evaluated in detail.
Results: 180 patients who had IEE were included in the study. 51% were females and 49% were males. The median age was 10 months (1month-7 years) and median body weight was 7.7 kg (3.3-61 kg). Patients were operated with the diagnosis of TOF (70 patients), VSD (50 patients), ASD (30 patients), complete AVSD (16 patients) and others cardiac diseases like mitral valve disease, subaortic ridge, pulmoner venous return anomaly (14 patients). Minor residual lesions were determined in 19.4% (n = 35) of patients without a need for re-entry to the cardiopulmonary bypass circulation (CPB). There were major residual lesions in 6.1% (n = 11) of patients that required re-entry to CPB. 4 of these major residua were in patients with VSD (large residua from the VSD patch), 4 with TOF (3 were severe right ventricular outflow tract stenosis, 1 was residua from VSD patch), 3 with complete AVSD (all were severe mitral regurgitation). No major complication was determined during IEE, only transient bradycardia was observed in 3 of the patients.
Conclusion: IEE provides a good guidance during congenital cardiac surgery in children. IEE helps to clarify the operative plan and helps to decrease morbidity and mortality due to unnecessary invasive procedures.

P2167 - CAN ABNORMALITIES OF THE MITRAL VALVE APPARATUS IDENTIFY GENOTYPE POSITIVE PHENOTYPE NEGATIVE FORM OF HYPERTROPHIC CARDIOMYOPATHY IN CHILDREN

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Background: Although described in adults with overt hypertrophic cardiomyopathy (HCM), abnormalities of mitral valve (MV) apparatus have been studied infrequently in children with HCM. Especially, little is known about MV in children with HCM gene mutations, without overt left ventricular (LV) hypertrophy (genotype positive, phenotype negative, GpPn). This study aims to describe two-dimensional echocardiographic (2DE) abnormalities of MV apparatus in GpPn children. We hypothesize that subtle variations in LV papillary muscles (PM) may be observed in GpPn children.
Methods: In this retrospective study, 2DE images of 27 GpPn and 27 control patients, were analyzed. Measurements included: apical displacement of PM base, angle between anterolateral PM and LV lateral wall, PM length, inter-PM distance at tips and area of PM in end-diastole and end-systole, length of MV leaflets in diastole, LV outflow tract dimension in systole. ANCOVA, multivariate logistic regression and ROC analysis were performed; a $p < 0.05$ was considered significant. Intraclass correlation (ICC) was applied.
Results: Of 27 GpPn children, 13 (48.15%) showed MYBPC3, 5 (18.52%) MYH7, 4 (14.81%) MYL2 mutations and 5 (18.52%) presented with other mutations. Echocardiographic characteristics are described in table 1. There was a significant increase in angle between anterolateral PM and LV lateral wall ($35.70° \pm 1.43$ vs. $28.43° \pm 1.43$, OR: 1.15 IC95%: 1.03–1.29), and decrease in inter-PM distance in end diastole (1.56 cm ± 0.07 vs. 1.84 cm ± 0.07, OR: 0.06 IC95%: 0.01–0.78) in the multivariate analysis. A cut point of > -0.3 was calculated for the multivariate model with 85.19% Sensitivity, 81.48% Specificity and 0.89 AUC (95% CI 0.80–0.98) (graph1), ICC: 0.966.

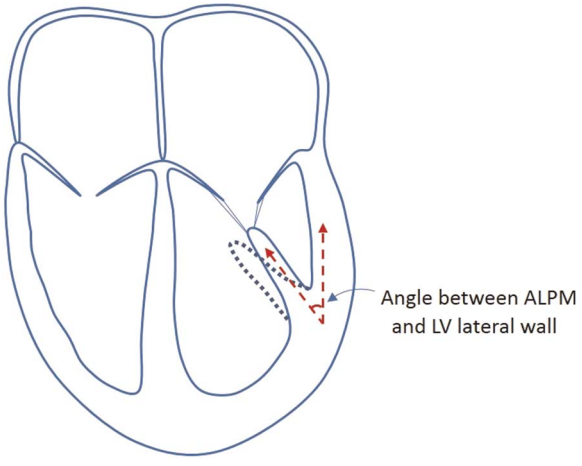

Figure 1.
Angle change in Genotype Positive, Phenotype Negative Patients

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Conclusions: GpPn children may manifest subtle abnormalities of MV apparatus. These echocardiographic parameters may serve as potential tools for identifying children with HCM gene mutations, without overt LV hypertrophy. Future multicenter, longitudinal studies may provide further insight.

Table 1. Characteristics of study population

	Controls (n = 27)	GpPn (n = 27)	p
Female n (%)	8 (29.63%)	13 (48.15%)	0.163
Age (years)	11.47 ± 0.44	10.69 ± 0.44	0.2468
IVSd (Z score)	-0.45 ± 0.20	0.03 ± 0.20	0.1181
LVPWd (Z score)	-0.28 ± 0.21	-0.53 ± 0.20	0.4491
Apical 4 Chamber view			
Anterolateral PM Apical Displacement (x/y)	0.50 ± 0.02	0.47 ± 0.02	0.2135
Anterolateral PM-LV lateral wall Angle (°)	28.43 ± 1.43	35.70 ± 1.43	0.0015*
Anterolateral PM Length (cm)	2.40 ± 0.10	2.24 ± 0.10	0.3066
Parasternal long axis view			
Anterior leaflet length (cm)	2.57 ± 0.08	2.67 ± 0.08	0.4039
Posterior leaflet length (cm)	1.09 ± 0.07	1.02 ± 0.07	0.5134
LVOT (cm)	1.76 ± 0.04	1.66 ± 0.04	0.0722
Short axis view			
Anterolateral PM Area (cm^2)	0.88 ± 0.07	1.04 ± 0.07	0.1572
Posteromedial PM Area (cm^2)	0.74 ± 0.08	1.15 ± 0.08	0.0013*
Inter PM distance Diastole (cm)	1.84 ± 0.07	1.56 ± 0.07	0.0082*
Inter PM distance Systole (cm)	0.78 ± 0.05	0.64 ± 0.05	0.068
LVOT (cm)	2.03 ± 0.08	1.90 ± 0.08	0.2899

IVSd: Interventricular septum at diastole. LVPWd: LV posterior wall at diastole

Apical displacement of PM was measured by dividing the distance of PM base to LV apex (x) with the distance from MV hinge point to LV apex (y).

Measurements were adjusted to body surface area (BSA), and are presented as adjusted means + SE

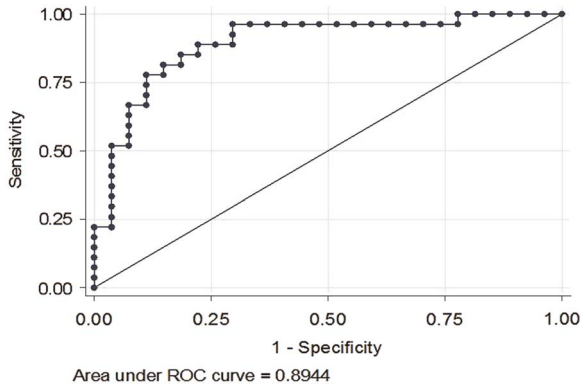

Figure 2.
ROC analysis.
*Equation: 3-2.80*Inter-Papillary muscle distance+0.14*PM angle-2.20*BSA Cut point of >-0.3 (85.19% Sensitivity, 81.48% Specificity)*

P2175 - ABNORMALITIES OF THE MITRAL VALVE APPARATUS IN CHILDREN WITH HYPERTROPHIC CARDIOMYOPATHY

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Background: Abnormalities of the mitral valve (MV) apparatus have been described in adults with hypertrophic cardiomyopathy (HCM). However, MV apparatus has been studied infrequently in children with HCM. We aim to outline two-dimensional echocardiographic (2DE) abnormalities of MV apparatus in children with HCM.

Methods: In this retrospective study, 2DE images of 35 HCM and 35 control patients, were analyzed. Measurements included: Apical displacement of base of anterolateral papillary muscle (ALPM), angle between ALPM and Left ventricular (LV) lateral wall, PM length, inter-PM distance at the tips in end diastole and end systole, MV leaflet length in diastole, LV outflow tract (LVOT) dimension in systole and area of PM base in diastole, both in short axis views. Student's T test was performed; a $p < 0.05$ was considered significant. Intraclass correlation (ICC) was assessed.

Results: Our cohort of 35 HCM children constituted a heterogeneous group of 29 (82.85%) non-syndromic HCM and 6 (17.14%) Friedreich's ataxia. Out of these non-syndromic HCM, 25.7% had obstructive HCM. The echocardiographic characteristics are described in table 1 and figure 1. The most important ones are 1) significant apical displacement of the base of PM 2) increase in the angle between ALPM and LV lateral wall, 3) elongation of entire ALPM, 4) elongation of MV leaflets and 5) decrease in inter-PM distance both in systole and diastole. ICC: 0.966.

Conclusions: Children with HCM manifest various MV apparatus abnormalities. In this descriptive study, the correlation between

Table 1. Characteristics of study population

	Controls (n = 35)	HCM (n = 35)	P
Female n (%)	10 (28.57%)	4 (11.43%)	0.330
BSA (m^2)	1.50 ± 0.30	1.60 ± 0.49	0.3079
Age (years)	13.66 ± 3.14	14.31 ± 5.03	0.0965
IVSd (Z score)	-0.26 ± 0.95	6.82 ± 5.51	<0.001
LVPWd (Z score)	-0.16 ± 1.09	3.25 ± 3.01	<0.001
Apical 4 Chamber view			
Anterolateral PM Apical Displacement (x/y)	0.49 ± 0.09	0.45 ± 0.08	0.0257*
Anterolateral PM-LV lateral wall Angle (°)	29.29 ± 5.04	35.72 ± 8.27	<0.001*
Anterolateral PM Length (cm)	2.56 ± 0.60	2.81 ± 0.72	0.111
Apical 2 Chamber view			
Anterolateral PM Apical Displacement (x/y)	0.59 ± 0.07	0.49 ± 0.08	<0.001*
Anterolateral PM-LV lateral wall Angle (°)	25.76 ± 7.71	31.07 ± 7.20	0.0041*
Anterolateral PM Length (cm)	2.19 ± 0.46	2.83 ± 0.80	<0.001*
Posteromedial PM Apical Displacement (x/y)	0.45 ± 0.10	0.35 ± 0.12	<0.001*
Posteromedial PM-LV lateral wall Angle (°)	28.72 ± 7.26	26.69 ± 10.31	0.1768
Posteromedial PM Length (cm)	2.49 ± 0.60	3.21 ± 1.02	<0.001*
Inter-PM distance in diastole	1.73 ± 0.53	1.43 ± 0.50	0.0175*
Inter-PM distance in systole	0.89 ± 0.33	0.52 ± 0.35	<0.001*
Parasternal long axis view			
Anterior leaflet length (cm)	2.75 ± 0.07	3.11 ± 0.11	0.0061*
Posterior leaflet length (cm)	1.12 ± 0.35	1.36 ± 0.40	0.0091*
LVOT (cm)	1.92 ± 0.04	1.95 ± 0.08	0.6875
Short axis view			
Anterolateral PM Area (cm^2)	1.14 ± 0.09	1.80 ± 0.13	<0.001*
Posteromedial PM Area (cm^2)	1.02 ± 0.40	1.51 ± 0.89	0.0677
Inter PM distance Diastole (cm)	2.00 ± 0.07	1.45 ± 0.08	<0.001*
Inter PM distance Systole (cm)	0.81 ± 0.05	0.50 ± 0.05	<0.001*
LVOT (cm)	2.24 ± 0.07	1.41 ± 0.10	<0.001*

IVSd: Interventricular septum at diastole. LVPWd: LV posterior wall at diastole

Apical displacement of PM was measured by dividing the distance of PM base to LV apex (x) with the distance from MV hinge point to LV apex (y).

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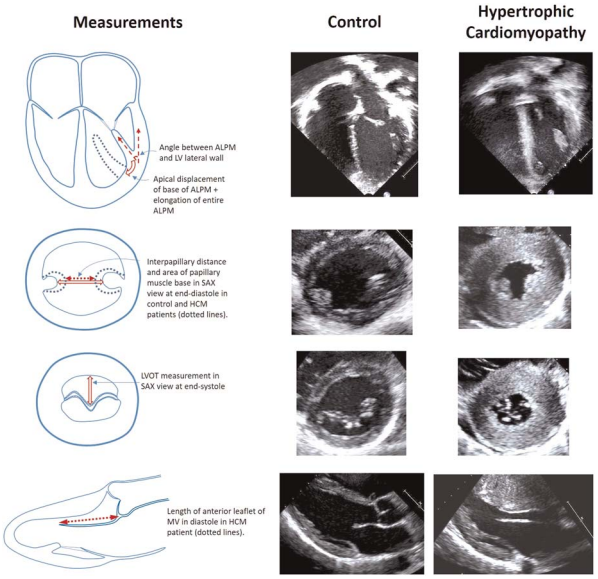

Figure 1.
Measurements of the Mitral valve apparatus

these parameters and clinical characteristics were not evaluated. We speculate that anomalies of MV may play a mechanistic role in LVOT obstruction and mitral regurgitation. Some MV abnormalities noted in this study may serve as adjunct diagnostic tools, in unusual cases of HCM. Further multicenter studies may offer a better understanding of the role of the MV apparatus in children with HCM.

P2177 - FETAL ECHOCARDIOGRAPHIC PREDICTORS FOR PERINATAL MORTALITY OF TRICUSPID VALVE MALFORMATION AND EPSTEIN ANOMALY

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Background: Tricuspid valve malformation and Ebstein anomaly (TVM/EA) diagnosed by fetal echocardiography shows huge clinical variety of severity and prognosis during perinatal period. Although Celermajer index and Simpson Andrews Sharland (SAS) score are well-established predicting scoring system for its prognosis, they do not reflect the hemodynamic impact of left ventricular function which is recently considered as more important in TVM/EA physiology. Therefore in this study, we aimed to clarify the predicting value of left ventricular function in TVM/EA fetuses.
Subjects and Methods: We reviewed the clinical records of 36 TVM/EA fetuses diagnosed between 2000 and 2015. We compared alive and dead cases by uni- and multi-variate analysis of the gestational age at diagnosis, cardiothoracic ratio (CTAR), direction of pulmonary flow and ductal flow, right-left ventricular ratio, tricuspid regurgitation (TR) velocity, Celermajer index, SAS score, and left ventricular (LV)-Tei index.
Results: Among 36 fetuses, two were terminated, one lost to follow-up, and two died in utero before 22 weeks of gestation. Of 31 fetuses, 12 were dead in perinatal period (perinatal mortality was 39%). The multivariate analysis revealed that TR velocity was significantly lower (3.2 ± 0.5 m/s vs 2.2 ± 0.5 m/s, $p < 0.01$) and SAS score was significantly higher (3.0 ± 0.5 points vs 6.0 ± 0.7 points, $p < 0.01$) in the dead cases. Absent pulmonary flow and retrograde ductus flow were also correlated with death. Importantly, LV-Tei index was significantly higher in dead cases (0.5 ± 0.05 vs 0.8 ± 0.08 $p < 0.01$). In contrast, there was no significant difference in gestational age, Celermajer index, CTAR, and right-left ventricular ratio. Finally we found that the combinational scoring system including TR velocity, pulmonary and ductal flow, and LV-Tei index could more efficiently predict the perinatal mortality of TVM/EA.
Conclusion: Our novel combinational scoring method including LV-Tei index is highly predictable for the perinatal mortality of TVM/EA. LV function might be a key regulator of prognosis in TVM/EA fetuses.

P2180 - COMPLICATION OR COINCIDENCE SUPERIOR VENA CAVA ANEURYSM AFTER SINGLE VENTRICLE PALLIATION OPERATION

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Background: Aneurysmal dilation of the SVC is a rare anomaly. Also it is ever rarer especially after single ventricle palliations like Glenn anastomosis or Fontan-Kreutzer procedure. Here in we report 5 casesof aneurysmal dilation of the SVC after single ventricle palliation.
Materials and Methods: Between January 20010 and November 2016 2950 cardiac catheterization procedure and more than 2584 congenital cardiac surgery was performed in our hospital. Among them, 75 patients had diagnostic cardiac catheterization before Fontan palliation for routine evaluation and 13 patients had cardiac catheterization after Fontan palliation for other reasons, 152 patients underwent single ventricle palliation (112 Glenn anastomosis and 40 Fontan). We detected 5 patients (2.2%) with SVC aneurysm.

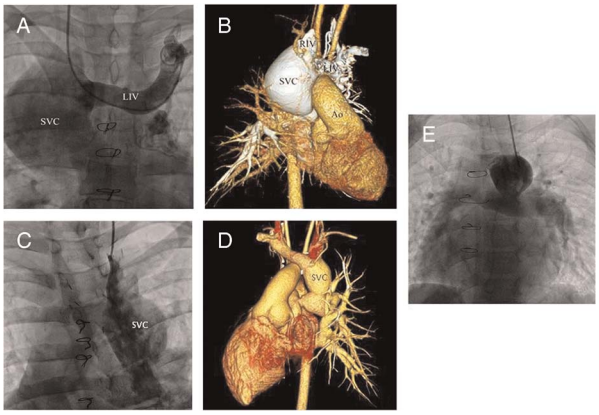

Figure 1.

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Results: (See Table-1)
Conclusion: The majority of the reported SVC aneurysms are either isolated pathology or secondary to connective tissue diseases. SVC aneurysms after single ventricle palliation is extremely rare and surprising. The common feature in all reported patients was the region of the aneurysm, yet we don't know why.

Table 1.

Patient	Sex	Age (years)	Cardiac pathology	Surgery	SVC aneurysm size {mm}	Mean PAP
1	Female	23	Double inlet left ventricle, rig ht vent ri cu 1 a r hy popl a si a, ventriculoarterial discordance	3years: Glenn 7 years: Extracardiac Fontan	57 × 81	Normal {6 mmHg}
2	Male	16	situs inversus dextrocardia, tricuspid atresia with ventriculoarterial discordance	10 years: Glenn	42 × 38	Normal (13 mmHg)
3	Female	9	Situs inversus levocardia. Left atrioventricular valve atresia Double outlet left ventricle with pulmonary stenosis	2,5 years: Glenn	34 x31	Normal (15 mmHg)
4	Male	12	Double inlet and double outlet undetermined ventricle with pulmonary stenosis	5 years: Glenn	40 x42	NA
5	Male	8.3	Double inlet and double outlet right ventricle with pulmonary banding	7 months: Pulmonary artery banding 3years: Glenn	34 × 51	Normal {14 mmHg}

P2190 - THE USE OF 3D TECHNOLOGIES FOR SURGICAL PLANNING OF COMPLEX CONGENITAL HEART DISEASE FIRST EXPERIENCE IN TURKEY

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Background: Double outlet right ventricle (DORV) and transposition of great arteries (TGA) are conotruncal anomalies with considerable heterogeneity of intracardiac anatomy. Echocardiographic imaging does not always provide the information needed to design an intracardiac baffle directing left ventricular out flow through the VSD to the aorta for biventricular repair. A three dimensional (3D) model of the heart based upon data derived from computed tomography (CT) may contribute to a more complete appreciation of the intracardiac anatomy. In this prospective study, we wanted to share our experience with surgical planning based on 3D modeling for complex intracardiac anatomy.
Methods: Based on CT images performed in selected patients with complex DORV and TGA anomalies, the intracardiac anatomy was segmented. Out of each patient's data set we generated a virtual computer model (3D mesh) and an STL-file that was printed as a 3D plaster model if necessary. Cuttings were made on the 3D mesh obtained to display the heart and associated anatomical structures were clarified.
Results: We printed two 3D models and created eight virtual models of patients with complex intracardiac anatomy and 4 of them were operated on (Table1). The models helped the pediatric cardiologists to better understand the spatial relationships between the ventricular septal defect and the great vessels, and the surgeons to identify risk structures, and to plan the surgical strategy in detail. There was a strong correlation between the 3D models of these patients and the anatomic state detected during the operation. However, information regarding valves, chordae and papillary muscles was insufficient in 3D models.
Conclusions: 3D cardiovascular models accurately replicate the patient's anatomy and are extremely helpful for planning surgery in complex intracardiac anatomy. In our experience, it enhances the alignment of pediatric cardiologists and surgeons by strengthening interdisciplinary team work. They may potentially reduce operative time and morbi-mortality.

Table 1. Patient Demographics, Diagnosis, and Surgical Outcome

Case	Age/Wei g ht(kg)	ECHO Diagnosis	Segmental anatomy	Past interventions
1	7 months, 5.5	DORV, D-malposed great arteries, severe PS with annularhypoplasia	S,L,D	PV perforation and RVOT stent
2	3 years, 12	DORV, anteroposterior great arteries, severe PS with annular hypoplasia	S,L,D	non
3	9 months, 10.5	DORV, D-mal posed great arteries, severe PS with mild annular hypoplasia	S,L,D	non
4	2 years, 9.7	TGA, VSD, anteroposterior great arteries, severe PS with annular hypoplasia	S,L,D	PDA stent
5	19 years, 45	TGA, VSD, situs inversus Lmalposed great arteries, severe PS with annularhypoplasia	l,L,L	BD Glenn shunt
6	3 years, 12.5	DORV with 1 arge i nl et VSD with si de-by-side great arteries (aorta right)	S,L,D	PAbanding
7	8 months, 7.3	TGA, VSD, D-mal posed great arteries, pulmonary atresia	S,L,D	Shunt, Systemicto pulmonary, Central
8	11 months, 4.8	TGA, D-mal posed great arteries, severe PS with mild annular hypoplasia	S,L,D	

Case	Additional diagnoses	Surgery Options	VSD position	Surgery Performed
1		BCPSorLVto aorta baffle and RVto PA conduit	remote	Biventri cular repair byLVto aorta baffle and RVto PA conduit
2	RVhypoplasia	BCPSorLVto aorta baffle and RVto PA conduit	remote	BCPS with
3	non	BCPSorLVto aorta baffle and RVOT procedure	Subaortic	Biventri cular repair byLVto aorta baffle and RVOTO repair*
4	LPA stenosis	BCPSorLVto aorta baffle and RVto PA conduit	Subaortic	Biventri cular repair byLVto aorta baffle and RVto PA conduit
5	Dekstrocardia	FontanorLVto aorta baffle and RVto PA conduit	Subaortic	Awaiting surgery
6		BCPSorLVto aorta baffle and RVto PA conduit	remote	Awaiting surgery
7		BCPSorLVto aorta baffle and RVto PA conduit	remote	Awaiting surgery
8	non	BCPSorLVto aorta baffle and RVto PA conduit	remote	Awaiting surgery

ECHO, Echocardiography; DORV, double outlet right ventricle; PV, pulmonary valve; BCPS, bidirectional cavapulmonary anastomosis; RVOT, right ventride outflow tract; RV, right ventricle; PA, pulmonary artery; LV, left ventricle; PS, pulmonary stenosis; RVOTO, right ventricle outf low tract obstruction; TGA, transposition of great arteries; VSD, ventricular septal defect; PDA, patent ductus arteriosus; LP A, left pulmonary artery. *(Ventriculotomy, Non-transanular patch)

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P2201 - INCREASE OF PROSTAGLANDIN E2 IN REVERSAL OF DUCTAL CONSTRICTION AFTER DIETARY RESTRICTION OF POLYPHENOLS

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Background/Hypothesis: It has already been demonstrated that substances with antiinflammatory effects which inhibit prostaglandin synthesis, such as nonsteroidal antiinflammatory drugs (NSAID) and polyphenol-rich foods, may cause fetal ductal constriction (DC). Reversal of DC after maternal restriction of polyphenols has been reported, but its relation to prostaglandin concentration has not yet been demonstrated. This study was performed to test the hypothesis that dietary intervention to decrease ingestion of polyphenol-rich foods improves DC and is followed by increase in prostaglandin E2 (PGE2) levels.
Material and Methods: Clinical trial with a healthy group as comparator. The interventional group (IG) was made up of third-trimester mothers whose single fetuses had DC, excluding those exposed to NSAID, and the control group (CG) only by third-trimester normal fetuses. The IG was submitted to dietary orientation to restrict polyphenol-rich foods and both groups answered a food frequency questionnaire after fetal Doppler-echocardiographic examination and blood draw for PGE2 levels analysis. After two weeks, the women were again submitted to fetal echocardiogram, dietary assessment and blood draw.
Results: Forty mothers were recruited for the CG and 35 for the IG. Mean maternal age (26.6 years) and mean BMI (30.12 kg/m2) were similar among the groups. In intragroup analysis, after dietary orientation the IG showed decrease in median polyphenol consumption (1234.82 to 21.03 mg/day, $p < 0.001$) and increase in median concentration of PGE2 (1091.80 to 1136.98 pg/ml, $p < 0.05$), in addition to normalization of Doppler signs of DC (decrease of mean systolic [1.92 to 1.34 m/s, $p < 0.001$] and diastolic [0.45 to 0.22 m/s, $p < 0.001$] velocities and increase in mean pulsatility index [1.96 to 2.53, $p < 0.001$]). In the same analysis, the CG did not show significant changes.
Conclusions: Dietary intervention to restrict polyphenol-rich foods in third trimester fetuses with ductal constriction increases prostaglandin E2 levels, with improvement of this condition.

P2239 - LV NON COMPACTION IN SUBJECTS WITH COARCTATION OF THE AORTA – PREVALENCE AND EFFECTS ON CARDIAC FUNCTION

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Background/Hypothesis: Left ventricular non-compaction (LVNC) has been associated with heart failure, arrhythmia, systemic thromboembolism and sudden cardiac death. CoA is one of the most common congenital lesions observed in a patient cohort with LVNC, however the prevalence of LVNC in patients with coarctation of the aorta (CoA) and its clinical significance remains unknown. We sought to evaluate the prevalence of LVNC in patients with repaired CoA as well as its effect on LV size and function.
Methods & Results: 268 patients (Mean age 26 (IQR 21-37) years, 63% male) undergoing cardiac MRI for clinical follow-up were included from three tertiary centres for adult congenital heart disease. Clinical data was obtained from medical records and correlated with ventricular volumes and function. LVNC was defined as a diastolic non-compacted:compacted dimension ratio >2.3 in the worst affected segment on a long-axis view as per current cardiac MRI criteria. The prevalence of LVNC was 8.2% (22 patients), with a mean end-diastolic NC:C ratio of 3.12 + /-0.8. The LV apical segment was affected in all 22 patients (100%), mid-segment in 11 patients (50%) and basal segment in 1 patient (5%). Indexed LV end-diastolic volumes (108 + /-32mL vs. 92 + /-22mL; $p = 0.006$) and stroke volumes (115 + /-28mL vs. 97 + /-24mL, $p = 0.003$) were significantly higher in the LVNC group compared to patients without LVNC. No significant differences in LV ejection fraction (56 + /-8% vs. 58 + /-11%; $p = 0.46$) were observed.
Conclusions: LVNC is relatively common in patients with repaired CoA and is associated with increased LV end-diastolic volumes and stroke volumes.

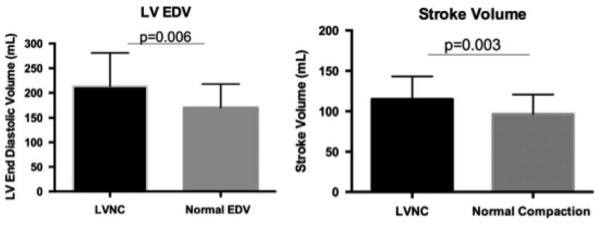

Figure.
LV Volumes in LV Non-compaction (LVNC) versus normally compacted controls, in patients with repaired coarctation.

P2272 - SIGNIFICANT DIAMETER HETEROGENEITY BETWEEN AORTIC ROOT AND AORTIC ANNULUS IN BICUSPID AORTIC VALVE

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Background: Bicuspid aortic valve (BAV) is associated with aortic dilatation and potential dissection. There are 3 major patterns of leaflets fusion in BAV: left and right coronary leaflets fusion (BAV-1), right and non-coronary leaflets fusion (BAV-2), and the rare three-leaflet fusion (BAV-3). We sought to determine the pattern of dilatation according to commonest types of BAV fusion.
Methods and Results: In this retrospective cross-sectional study, echocardiography reports were reviewed to evaluate the homogeneity of normalized diameters of the aortic valve, the aortic root and the ascending aorta according to fusion morphology. From 216 BAV patients followed at CHU Sainte-Justine between 2009 and 2014 (mean age at echocardiography 1 was 7.19 ± 5.56 years; 74.5% males), 155/216 (71.8%) had BAV-1, and 61/216 (28.2%) had BAV-2; $p = NS$ for age. Aortic valve stenosis was present in 66/216 (30.6%) patients, with a lower prevalence 39/155 (25.2%) in BAV-1 compared to 27/61 (44.3%) in BAV-2 ($p = 0.006$). There was no statistically significant difference in the ascending aorta between the study groups (Table 1). However, BAV-1 had increased aortic root Z-score compared to BAV-2 ($p = 0.0007$) despite a trend towards larger aortic annulus in BAV-2 ($p = 0.11$). The difference was not significant between BAV-1 and BAV-2 in cases with valvular stenosis (0.67 ± 1.50 vs. 0.17 ± 1.73; $p = 0.48$), but significant in the absence of stenosis (0.79 ± 1.57 vs. -0.25 ± 0.98; $p = 0.00035$). Altogether, there was no significant differences in the ascending aorta dimension in either case (BAV-1 vs. BAV-2, or valvular stenosis vs. no stenosis).

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Conclusion: Although not necessarily dilated (mean Z < 2.0), type-1 BAV fusion aortic roots are larger than in type-2. The observed difference suggests potential modifications in patient approach with this valvular disease according to the type of fusion.

Table 1. Z-scores based on fusion type (mean ± SD)

	BAV-1	BAV-2	p value
Aortic Valve (AV)	0.43 ± 1.71	0.88 ± 1.90	0.11
Aortic Root (AR)	0.70 ± 1.50	-0.08 ± 1.34	0.0007
Ascending Aorta (AA)	1.45 ± 2.13	2.04 ± 2.15	0.07

P2315 - TYPES AND OUTCOMES OF CONGENITAL CARDIAC MALFORMATION IN INDONESIAN PATIENTS WITH ISOMERIC HEART

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Background: Isomerism or Heterotaxy syndromes are rare congenital anomalies which has high morbidity and mortality. This term includes patients with a wide variety of very complex cardiac lesions. The hallmark of this cardiac anatomy includes isomeric right or left atria, which can be diagnosed by echocardiography.
Objectives: To describe the types of congenital cardiac malformation in patients with isomeric heart based on echocardiography and its outcomes.
Methods: We reviewed all patients with isomeric heart who were assessed and managed at National Cardiovascular Center Harapan Kita, Jakarta, Indonesia from January to December 2016. Isomeric heart was diagnosed by echocardiography and most of them had undergone cardiac surgery.
Results: There were 54 patients with isomeric heart with age ranged from 10 days to 27 years old. Thirty four (63%) were patients with right isomeric heart, 20 (37%) with left isomeric heart. The overall most common cardiac anomalies were atrio-ventricular septal defect in 38 (70%), double outlet right ventricle in 19 (35%), pulmonary atresia in 15 (27%), and total anomalous pulmonary venous drainage in 12 (22%) patients. Surgery was performed in 32 (66%) patients age ranged from 12 days to 13 years old with hospital mortality 9 (16%) patients. The most common cause of death of these patients were severe infection 2 (22%), arrhythmia 1 (11%), and additional airway anomaly 1 (11%). Five (9%) patients died before surgery due to infection, severe hypoxia, gastro-intestinal problems or airway problems. From all 14 deaths, 6 (43%) patients were neonates and 7 (50%) had extra cardiac total anomalous pulmonary venous drainage.
Conclusion: The most common cardiac anomaly found in our isomeric heart patients was atrio-ventricular septal defect. The early outcome of these patients is still not satisfactory despite of better medical facilities and surgical management.

P2323 - FUNCTIONAL PRENATAL ECHOCARDIOGRAPHIC PATTERN AS A PREDICTOR OF SEVERITY OF POSTNATAL AORTIC COARCTATION

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Background: Coarctation of the aorta (CA) is a common form of congenital heart defect which may require an intervention during infancy. Our main goal was to identify an echocardiographic prenatal ventricular functional pattern associated with the severity of CA.
Materials and Methods: Retrospective review of patients with suspected CA with prenatal functional echocardiography, which diagnosis was postnatally confirmed between June 2011 and September 2016 in our institution. Comprehensive echocardiography was performed at third trimester of pregnancy. Biventricular systolic and diastolic functions were assessed by M-mode, pulse-wave and tissue Doppler imaging (TDI techniques). Postnatal transthoracic echocardiographic was obtained to measure the aortic valve, ascending aorta and the transvers and isthmic arch. Z-scores were calculated. Clinical courses were also reviewed. Statistical analysis comparing functional prenatal parameters with the size and the z-scores of the aorta was conducted.
Results: Twenty one patients were admitted to the NICU and 20/21 patients received prostaglandin infusion. All interventions were performed between 2 and 12 days of life (3/20 balloon angioplasty, 8/20 end-to-end repair, extended arch repair 7/20 and Norwood approach 1/20) except in one case which underwent balloon angioplasty at 103 days of life. A negative correlation between prenatal Pulmonic Valve size and postnatal Transverse arch diameter ($r = -.55$, $p < 0.01$) and positive correlation between Right End-Systolic Sphericity and Isthmic diameter ($r = .81$, $p < 0.01$) was found. Those patients undergoing extensive arch repair/Norwood operation had higher prenatal Right Cardiac Output (RCO) compare to those undergoing Angioplasty/end-to-end repair (0.6 ± 0.3 vs 0.9 ± 0.3, $p = 0.04$). No other significant correlations between echocardiographic prenatal ventricular functional pattern and the severity of CA were found.
Conclusions: In our study patients requiring extensive arch repair had significantly higher prenatal RCO compared with those requiring simple arch reconstruction. Larger series are needed to assess whether there is a functional parameter that could better predict the postnatal severity of the CA.

P2332 - ALTERATION IN CEREBRAL BLOOD FLOW IN CHILDREN WITH CONGESTIVE HEART FAILURE DUE TO VENTRICULAR SEPTAL DEFECT

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Aim: We aimed to investigate the effect of ventricular septal defect (VSD) and heart failure on cerebral blood flow (CBF) and whether there is any improvement in CBF after the treatment of heart failure and any relationship with serum NT-proBNP.
Method: Forty children with VSD (13 of them had heart failure) aged between 1-36 months were included in the study group. Control group consisted of 25 healthy children at age of 1-36 months. Maximum, minimum and mean blood flow velocities and pulsatile indexes of right and left medial cerebral arteries were assessed through the temporal window by transcranial Doppler ultrasonography. CBF measurement and serum NT-proBNP

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analysis were repeated in patients with heart failure a month after the beginning of anticongestive therapy. All cases were scored and grouped according to the Ross scoring system. The groups were also compared in terms of defect diameters, cardiac structural changes, left ventricular systolic functions and the findings related with pulmonary hypertension determined by echocardiography. The correlations between echocardiographic measurements and CBF parameters were analyzed.
Results: Although no difference was found between the patient and control groups for CBF ($p > 0.05$), right and left maximum CBF velocities significantly increased after anticongestive therapy in patients with heart failure ($p = 0.04$; $p = 0.01$, respectively). NT-proBNP level of patients with heart failure was significantly higher than the patients with VSD without heart failure ($p = 0.04$) and control groups ($p < 0.001$). NT- proBNP levels negatively correlated with right and left maximum CBF velocities ($r = -0.39$, $p = 0.013$; $r = -0.32$, $p = 0.043$, respectively).
Conclusion: Although no difference was found among study groups for CBF velocity, occurrence of an increase in CBF velocity after anticongestive therapy and negative correlations between CBF velocity and both the defect diameter and NT- proBNP levels have shown that hemodynamic status due to VSD and heart failure has an effect on CBF.

P2339 - RISK FACTORS FOR PROGRESSIVE AORTIC DILATATION IN CHILDREN WITH BICUSPID AORTIC VALVE (DATA FROM REVAB)

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Objectives: Define risk factors for developing progressive aortic dilatation in children with bicuspid aortic valve.
Methods: A total of 393/944 patients had 2 visits introduced in the Spanish registry for children with bicuspid aortic valve (REVAB), from the Spanish Society of Pediatric Cardiology and Congenital Heart Diseases (SECPCC). Only patients with at least 1 year between visits were included. Progression was defined as standard deviations adjusted by weight and height per year followed.
Results: Mean progression of sinus of Valsalva was 0.21 ± 0.59 SD/y and was related with younger age at both first and second visit -3.12, $p = 0.002$ and -3.87, $p < 0.001$, and concomitant coarctation of the aorta 3.08, $p = 0.002$. However, patients with aortic regurgitation had less progression -1.99, $p = 0.047$. No association was found with aortic stenosis -0.053, $p = 0.596$, morphology of the aortic valve (pure 1.56, $p = 0.12$, horizontal -1.69, $p = 0.09$) or time of follow-up -1.89, $p = 0.06$. In the multivariate analysis, only patients who were younger at second visit and with concomitant coarctation of the aorta remained associated with progression of the aortic root dilatation, -3.52 $p = 0.001$ and 2.77 $p = 0.006$ respectively. Mean progression of ascending aorta was 0.29 ± 0.67 SD/y and was related with younger age at second visit -2.74, $p = 0.007$, left sided obstructive lesions 2.36, $p = 0.019$, and shorter follow-up -2.55, $p = 0.011$. There was no association with age at first visit -1.75, $p = 0.08$, aortic stenosis -0.27, $p = 0.788$, aortic regurgitation -1.17, $p = 0.242$, or morphology of the aortic valve (pure -0.98, $p = 0.327$, horizontal 1.71, $p = 0.088$). In the multivariate analysis, patients younger at second visit, with concomitant left sided obstructive lesions and shorter follow-up remained associated with progression of the ascending aorta dilatation: -2.04 $p = 0.04$, 2.19 $p = 0.03$ and -2.08 $p = 0.039$ respectively.
Conculsion: Younger patients with concomitant coarctation of the aorta or other left sided obstructive lesions are at higher risk of progression of the aortic dimensions.

P2341 - DIFFERENT PATTERNS OF AORTIC DIMENSIONS IN CHILDREN WITH BICUSPID AORTIC VALVE (DATA FROM REVAB)

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Objectives: Define age-wise patterns of aortic dimensions among children with bicuspid aortic valve.
Methods: From the Spanish registry for children (<18 years) with bicuspid aortic valve (REVAB), under the auspices of Spanish Society of Pediatric Cardiology and Congenital Heart Diseases (SECPCC), patients with isolated bicuspid aortic valve (589) were compared to patients with concomitant congenital anomaly (including coarctation

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of the aorta, interrupted aortic arch, subaortic stenosis, mitral stenosis, ventricular septal defect and others, 355). Dimensions of the aorta were adjusted by weight and height (standard deviations, SD).
Results: Overall, patients with isolated bicuspid aortic valve had larger dimensions of the aortic root and the ascending aorta: 0.2 ± 1.6 SD vs -0.1 ± 1.9 SD and 1.5 ± 1.9 SD vs 0.4 ± 2.4 SD, $p = 0.005$ and $p < 0.001$ respectively. Age-wise aortic dimensions differed between the groups (see figures attached). Patients with isolated bicuspid aortic valve (green) had larger sizes of the aortic root at birth with a decrease until 6 years of age and then stabilized; the ascending aorta was also larger, with a mean over 2 SD, at birth and started growing even more between 6 and 10 years of age, and then stabilized. Patients with concomitant congenital disease (violet) had smaller sizes of the aortic root at birth with an increase until 5 years of age, then stabilized and after 12 years of age started increasing again. Similarly, patients with concomitant congenital disease had smaller ascending aorta at birth, with an increase until normalization or even slightly dilatation with a peak at 8 years of age and then stabilized.
Conclusions: Patients with isolated bicuspid aortic valve have larger aortic dimensions at birth with little change over somatic growth. On the other hand, patients with concomitant congenital anomaly have smaller aortic sizes at birth with a trend towards progressive dilatation over somatic growth.

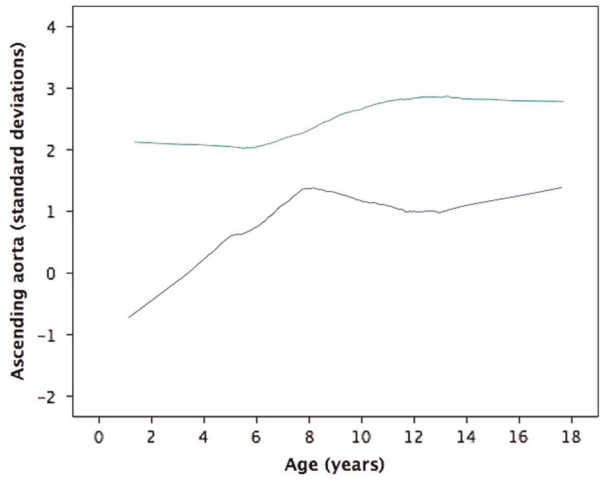

Figure 1.

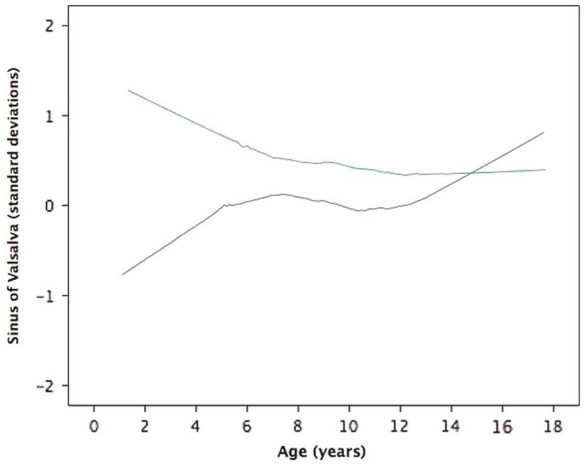

Figure 2.

P2342 - NATIVE T1 MAPPING OF LEFT VENTRICLE OF CHILDREN WITH POST CORONARY RELATED CARDIAC SURGERY

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Background/Hypothesis: Myocardial fibrosis is a major independent predictor of adverse cardiac outcomes in children with congenital heart diseases. T1 mapping of left ventricular (LV) wall using Cardiac Magnetic Resonance Imaging (CMR) has emerged as an alternative technique in detection of fibrosis. Primary objective of this study is to ascertain T1 mapping values of children with congenital heart diseases where the primary corrective surgery involved coronary arteries re-implantation/transfer. This cohort was hypothesized to have early fibrosis evidence with T1 parameters which are higher than controls.
Methods: 25 paediatric patients (mean age 12.7 + /- 3.8 years old) who either had arterial switch Operation/Anomalous Left Coronary Artery from Pulmonary Artery (ALCAPA) repair/Ross procedure/double switch operation were included in the study. All had undergone CMR from March-December 2016 at Birmingham Children's Hospital, Birmingham, United Kingdom. A single breath-hold Modified Look-Locker Inversion-recovery (MOLLI) pulse sequence was used on a mid-ventricular short axis imaging plane. 27 patients were also included as control.
Results: All patients in the cases group had normal LV function (mean LVEF : $65.8 \pm 10.8\%$, LVEDVi 68.3 ± 11.4 ml.m2, SVi 44.4 ± 8.8 ml.m2). In comparison of native T1 values of inter-ventricular septum, no significance difference were found between case cohort and control (982.7 ± 36.8 vs 979.7 ± 29.0, $p = 0.74$). Native T1 values of LV free walls were also not statistically different between the two groups (954.7 ± 41.1 vs 956.6 ± 24.9, $p = 0.84$). However in the cases cohort, T1 of LV free wall are lower than inter-ventricular septum (954.1 ± 41.4 vs 982.7 ± 36.8, $p = 0.015$).
Conclusions: Native T1 values are not significantly higher in children who had cardiac surgery with coronary re-implantation/transfer and normal post-surgical left ventricular function. However, further study needs to be performed in the cohort with impaired left ventricular function.

P2348 - EXTRA CELLULAR VOLUME IMAGING OF LEFT VENTRICULAR WALLS IN CHILDREN WITH CONGENITAL HEART DISEASES AND IMPAIRED VENTRICULAR FUNCTION

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Background/Hypothesis: In children with congenital heart diseases, myocardial fibrosis is a possible long term complication with impairment of left ventricular function. Extra-Cellular Volume (ECV) imaging of left ventricular (LV) walls using Cardiac Magnetic Resonance Imaging (CMR) offers early detection of fibrosis before late gadolineum enhancement (LGE) changes are seen. Aim of the study was to determine ECV of a cohort of children with congenital heart diseases and any association with their left ventricular function.

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Materials and Methods: 19 children with congenital cardiac conditions who had undergone CMR from March to December 2016 at Birmingham Children's Hospital UK were included in the study. All subjects underwent CMR (Siemens Avanto 1.5 T scanner) to assess LV function, measurement of ECV on T1-mapping (MOLLI sequence) and standard late gadolinium enhancement (LGE) imaging. ECV values were determined from 5 different LV wall segments on short axis images (inter-ventricular septum, anterior, antero-lateral, inferio-lateral, inferior segments).
Results: 4 children (age 10.9 ± 3.9 years old) had impaired LV ejection fraction (44.5 ± 6.0%) and increased LV end diastolic volume indexed (82.2 ± 14.5 ml.m2). One of the children had LGE changes seen on the inter-ventricular septum. ECV parameters of the inter-ventricular septum were higher in children with impaired LV function (38.6 ± 7.0% vs 30.0 ± 3.5%, $p = 0.002$). No significant difference were found in the ECV of LV free walls (32.2 ± 5.2% vs 28.7 ± 5.7%, $p = 0.279$).
Conclusions: ECV technique has promising possibility in detection of myocardial fibrosis in children with impaired LV function, similar as adult studies. In those with poor LV function, there is a strong possibility that area of fibrosis is in the inter-ventricular septum. However, normal ECV parameters will need to be determined first in children before comparing with those with cardiac illnesses.

P2349 - THE TREND CHANGE IN CARDIAC IMAGING IN CONGENITAL HEART DISEASE EXPERIENCE OF TERTIARY CENTER IN INDONESIA

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Background: Cardiac imaging is fundamental for diagnosing and evaluating of congenital heart disease (CHD). Transthoracic echocardiography (TTE) as the first-line imaging modality in CHD, has some limitations. The use of cardiac computed tomography (CT) and cardiac magnetic resonance imaging (CMRI) as further non-invasive diagnostic modalities are now being reconsidered instead of invasive diagnostic cardiac characterization.
Objective: To evaluate the change of cardiac imaging for CHD at National Cardiovascular Center Harapan Kita Jakarta, Indonesia.
Method: Data was obtained through a retrospective manner over 4 years from diagnostic medical records of CHD patients from 2013-2016.
Result: During the study period, an average of 8182 diagnostic tests were performed annually. The number of total studies TTE, diagnostic cardiac catheterization, Cardiac CT and CMRI procedures change significantly over time. The average use of TTE, diagnostic cardiac catheterization, cardiac CT and CMRI annually are 7089 cases, 452 cases, 167 cases and 473 cases per-year respectively. The use of cardiac MRI (0.83%, $p = 0.01$), and cardiac CT (3.77%, $p < 0.072$) increased linearly. Whereas that the use of echocardiography (88.9%, $p = 0.005$) and invasive diagnostic catheterization decreased (6.49%, $p = 0.458$) with relatively stable number of non-surgical CHD intervention (45.71%, $p = 0.844$). Cardiac CT evaluation is mostly performed in younger patients whom require evaluation of anatomy, great artery and anomaly of vein. Cardiac MRI examination was especially performed in older patient whom require anatomy, tissue characterizations, volume, function, and flow measurement. Invasive diagnostic catheterization was mostly performed among patients whom require hemodynamic evaluation.
Conclusion: Diagnostic imaging on CHD underwent changes due to the development of the use of CMRI and Cardiac CT. The choice of further imaging modalities following echocardiography examination depends on the clinical questions that remain to be addressed.

P2352 - EARLY ATHEROSCLEROTIC CHANGES AND MYOCARDIAL FUNCTIONAL ALTERATIONS IN CHILDREN WITH ISOLATED BICUSPIT AORTIC VALVE

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Objectives: Bicuspid aortic valve (BAV) is the commonest congenital cardiac malformation with a prevalence of % 0,5 to 2%. It is frequently associated with dilatation, aneurysm and dissection of the ascending aorta. The purpose of the following study is to evaluate function of the left ventricle and stiffness, elasticity and strain of ascending aorta and epi-aortic vessels wall and thickness of the carotis intima media in pediatric patients with BAV.
Methods: Twenty four pediatric BAV patients (mean age 11.7 ±3.1 years) with no or mild valvuler impairment and 20 healty control subjects (mean age 11.9 ± 3.1) were included in the study. Global myocardial functions were evaluated with tissue Doppler derived myocardial performance index. We assessed the mean and maximal values of carotid intima-media thickness (CIMT) of the carotid artery. Aortic and carotid strains, distensibilities and stiffness indexes were derived.
Results: Diastolic blood pressure values of BAVs were higher than controls ($p < 0.001$). Diastolik and systolic diamaters of assendan aorta and sinotubuler junction and sinüsvalsalva diameters were higher than the control group (respectively <0.001, <0.001, $p < 0.05$, $p < 0.04$). Maximal values of CIMT of bilateral carotid arteries were higher than controls (respectively $p = 0.006$, $p = 0.003$). Isovolumetric contraction time, isovolumetric relaxation time and myocardial performance index obtained with tissue Doppler echocardiography were higher in BAVs (respectively $p = 0.016$, $p < 0.001$, $p < 0.01$). No significant differences were found in elastic properties of aorta and carotid arteries. . There was positive correlation between CIMT and isovolumetric relaxation time, diastolic and systolic blood pressure and diastolic diameter of assendan aorta.
Conclusions: In patients with isolated BAV, the blood pressure profile was elevated and was associated with left ventricular dysfunction and early atherosclerotic changes.

P2355 - EVALUATION OF MYOCARDIAL RESERVE WITH LOW DOSE DOBUTAMINE STRESS ECHOCARDIOGRAPHY IN PATIENTS WITH SEVERE LEFT ATRIOVENTRICULAR VALVE REGURGITATION IN THE LATE POSTOPERATIVE PERIOD OF ATRIOVENTRICULAR SEPTAL DEFECT REPAIR

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Background: The management of oligosymptomatic patients with severe left atrioventricular valve (LAVV) regurgitation in the late follow-up of atrioventricular septal defect (AVSD) repair is still challenging. Surgical repair follows the echocardiographic guidelines for mitral valve, where surgery is suggested when there is left ventricular (LV) dysfunction or increased LV systolic diameter. However, ejection fraction (EF) remains preserved for a long period of time, despite of the grade of mitral valve insufficiency. Dobutamine stress echocardiography (DSE) has shown to be a sensitive test for detection of subclinical cardiac dysfunction and can potentially assess functional myocardial reserve (MR). This study aimed to evaluate MR of oligosymptomatic patients with severe LAVV regurgitation in the late follow-up after AVSD repair.
Methods: 27 oligosymptomatic outpatients (18.6 ± 12.8 years-old) were studied. EF (Simpson's rule) and LV Global Longitudinal Strain (GLS) were measured at rest and after 10 minutes of dobutamine infusion (5 mcg/kg/min). Preserved MR was considered when EF increased at least 5% or the resting value.
Results: 18 patients had partial AVSD, 9 had the total form. At rest, mean EF was 64 ± 4.2% (55-72%) and GLS 20.9 ± 2.6% (15.9-26.4%). During stress, EF increased to 74.4 ± 7.1% (59-85%), GLS to 24.2 ± 2.9% (17.1-30.4%). 3 patients (11%) had no MR. In those, increment on GLS at stress was significantly lower than in preserved MR group (17.3 ± 1.07% to 18.2 ± 1.14% vs 21.4 ± 2.37% to 24.9 ± 2.1%; p:0.005). A cut-off point of 1.5% increment in GLS during stress was identified as preserved MR (Kappa index: 1).
Conclusion: This study detected the occurrence of 11% of absent MR in this group, which reflects the inadequate heart responses to stressing life situations and, therefore, it turns earlier the moment of surgery. The increment of LV GLS less than 1.5% after Dobutamine infusion was related to absent myocardial reserve.

P2365 - FREQUENCY OF CARDIAC PATHOLOGY IN FETAL ECHOCARDIOGRAPHY

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In this study; a frequency of cardiac pathology was examined in patients who were sent to the Pediatric Cardiology Clinic for fetal echocardiography.
Material and Method: In this study; the results of 839 patients who had fetal echocardiography in the pediatric cardiology clinic between 2009-2016 have been examined retrospectively using Microsoft Excel 2010.
Results: Within the study 818 fetuses of 839 mothers have been examined. 19 cases were multiple pregnancy, whereas in 4 cases examination was done for two separate fetuses. 85% of the cases were patients from our hospital's obstetric clinic, the remaining from other clinics. The average age of the mothers was 30 (17-55). The average gestation week was 25 (18-37). When the reason for fetal echocardiography was examined it was found out that 29.22% didn't have a reason,18.95% had maternal reasons, 32.89% had fetal reasons, 5.38% had familial reasons and the rest had multiple reasons. In 17.18% of the cases cardiac pathology was detected during fetal echocardiography. 2.86% of the cases had serious cardiac anomalies. Hyper echogenic focus was seen in 8.46% of the cases. Only 34.68% had newborn echocardiography in our clinic; and the cardiac pathology percentage was 16.15%. In 5.31% of the cases where cardiac pathology was seen in fetal echocardiography, the anomaly wasn't witnessed later on during newborn echocardiography. In 29.71% cases that weren't diagnosed with cardiac pathology in fetal echocardiography, cardiac pathology was seen in newborn echocardiography.
Conclusion: Fetal echocardiography is mostly a reliable, non-invasive method for diagnosing cardiac pathology. However, in some cases it was seen that cardiac pathology was seen in newborn echocardiography when it wasn't seen in fetal echocardiography; and vice versa. This shows that although fetal echocardiography gives accurate results in many cases, it isn't safe to assume that it is a reliable method in each case.

P2379 - PRENATAL RINGS DIAGNOSED IN PRENATAL TIME. CORRELATION WITH POSTNATAL IMAGE ON MDCT

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Background: Vascular rings (VR) are congenital abnormalities of the aortic arch consisting of vascular structures that encircle and sometimes compress the airway and esophagus. The most frequent complete VR are double aortic arch (DAA) and the right arch with anomalous left subclavian (RAA + ALSA) and left persistent ductus. In recent years, with prenatal diagnosis, it has been an increased frequency of VR with respect to as described in postnatal series, demonstrating greater percentage of asymptomatic patients during follow-up. Multidector computed tomography (MDCT) is the standard gold test for VR study. Our main objective has been to study the concordance of prenatal ultrasound on the MDCT findings.
Materials and Methods: Retrospective study of patients with prenatal diagnosis of VR in La Paz Children's Hospital between 2010-2016 which performed in postnatal life MTCD.
Results: In total, we had 19 patients (12 patients diagnosed AAD and 7 patients RAA + ALSA). 100% of patients who were diagnosed of VR by fetal echocardiography, the existence of a ring was confirmed by MCTD. Regarding the type and morphology of ring, only in one case was defined in prenatal ultrasound the RAA + ALSA, described in MTCD as AAD + suspended left subclavian artery. 36% of patients presented secondary symptoms to ring (57% of these secondary to DAA), being the most frequent respiratory symptoms. 100% of symptomatic patients required surgery (78% first year of life). Surgery of the ring was performed in two asymptomatic patients during a surgical procedure for another heart disease.
Conclusions: After the incorporation of the 3-vessel and tracheal views, the detection of VR in prenatal period has increased the frequency and improved knowledge of natural history. The concordance in our hospital between the diagnosis of VR by fetal echocardiography respect to MDCT is very high (94.7%), allowing a great precision prognostic.

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P2382 - EVALUATION OF HOME FETAL HEART RATE DOPPLER MONITORING FOR SURVEILLANCE OF FETAL ARRHYTHMIAS

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Background: Standard of care (SOC) for outpatient monitoring of fetal arrhythmias (FA) includes assessments in obstetrical office 2-3 times/wk. plus frequent fetal echocardiograms. At Johns Hopkins All Children's Hospital (JHACH), this SOC was changed to add home monitoring for 1-2 min 3 times/day with a fetal heart rate monitor (FHRM) which was provided to mom carrying babies with FA. The study's primary objective was to determine concordance of the FHR's obtained with both methods, home FHRM and Doppler ultrasound (DU) at the office. Secondary objectives are to compare time to diagnosis, time to treatment, and outcomes of fetuses monitored with the FHRM as compared to the previous SOC.
Methods: A retrospective cohort study was implemented (Table 1). We studied two cohorts: Cohort 1: (Jan 2008-May 2013, previous SOC), and Cohort 2: (May 2013- October 2016, modified SOC). FHR data obtained with FHRM and the FHR obtained by DU after the second visit were compared for agreement/concordance. Secondary objectives and prenatal and postnatal survival will be compared between the cohorts in upcoming analysis.
Results: For cohort 2, data on 13 subjects was available. Bland-Altman analysis (Figure 1) showed that the mean difference in FHR between the FHM and DU was -0.27 (95% CI = -2.87-2.33). The limits of agreement (+2 SD of the mean) ranged from -8.89- + 8.35. Barring 1 measurement, all the other measurements had good agreement between the paired measures. The concordance correlation coefficient was 0.98 (95% CI = 0.96-0.99), indicating strong agreement. Good compliance with outpatient monitoring was noted and there was no evidence of perinatal complications.
Conclusions: There was strong agreement in FHR between the measures obtained with the home monitors and the Doppler ultrasound. Based on these preliminary results, the addition of adjunctive home FHR monitors to the SOC appears to be feasible and likely reliable.

Table 1. Study design/ Timeline

Old SOC	New SOC	Concordance FHR	Log Book Data	Fetal Echo Data	Time to diagnosis/ treatment	Survival/ Postnatal Outcomes
Cohort 1 Retrospective 1/1/2008-5/31/2013				X	X	X
	Cohort 2 Retrospective 6/1/2013-9/21/2016	X	X	X	X	X

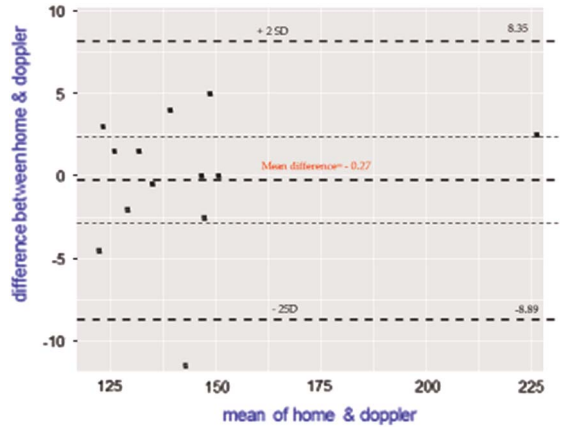

Figure 1.
Brand-Altman analysis of agreement between FHR monitors and DU (Cohort 2)

P2392 - IMPACT OF FIRST TRIMESTER CARDIAC SCREENING ON UN NATURAL HISTORY OF CONGENITAL HEART DEFECTS

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Background: The study analyzed the impact of first trimester screening on the spectrum of congenital heart defects (CHD) later in pregnancy, and on the outcome of fetuses and children born alive with CHD.
Materials and Methods: The spectrum of CHD, associated comorbidities (chromosomal and/or structural non-cardiac anomalies) and outcome of fetuses either diagnosed with CHD by echocardiography in the first trimester (Group I, 127 fetuses), or only in the second trimester screening (Group II, 344 fetuses), were analyzed retrospectively between 2007 and 2013. Second trimester fetuses diagnosed with CHD between 2007 and 2013 were also compared with Group III (532 fetuses diagnosed with CHD in the second trimester from 1996 to 2001, the period before first trimester screening was introduced).
Results: The spectrum of CHDs diagnosed in the first and second trimesters in the same time period (from 2007 to 2013) differed significantly, with a greater number of comorbidities (chromosomal and/or structural non-cardiac anomalies) ($p < 0.0001$), CHDs with univentricular outcome ($p < 0.0001$), intrauterine deaths ($p = 0.01$) and terminations of pregnancy ($p < 0.0001$) in Group I in comparison to Group II. In Group III, significantly more cases of CHD with univentricular outcome ($p < 0.0001$), intrauterine demise ($p = 0.036$) and early termination ($p < 0.0001$) were identified in comparison to fetuses diagnosed with CHD in second trimester between 2007 and 2013. The spectrum of CHD seen in the second trimester groups differed after first trimester screening was implemented.
Conclusions: First trimester screening had a significant impact on the spectrum of CHD and on the outcomes of pregnancies with CHD diagnosed in the second trimester. Early detection of severe forms of CHD and significant comorbidities resulted in an increased pregnancy termination rate in the first trimester.

P2408 - USEFULNESS OF THREE DIMENSIONAL ECHOCARDIOGRAPHY FOR THE ASSESSMENT OF RIGHT VENTRICULAR FUNCTION IN CHILDREN WITH ARRHYTHMIA – A PRELIMINARY REPORT

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The accurate assessment of right ventricular function in children is still a diagnostic challenge. Cardiovascular magnetic resonance (CMR) is considered the gold standard in assessing RV function. Majority of arrhythmic disorders in children are classified as idiopathic as no link to abnormal cardiac morphology or function can be found. The aim of the study was to assess right ventricular function in children with arrhythmia using three-dimensional echocardiography and compare it to two-dimensional echocardiography and cardiac magnetic resonance.
Methods: The study group consisted of 18 children aged 6-17 years, average 13,5 ± 2,7 years, with arrhythmic disorders: 13 with ventricular arrhythmia (5 with single ectopic beats, 8 with complex arrhythmia) and 5 with supraventricular arrhythmia. In all children 2D and 3D echocardiography and in 7 also CMR were performed. The right ventricular ejection fraction (EF) was assessed and compared using Spearman correlation test.
Results: 15 out 18 patients had normal heart morphology while in 3 arrhythmogenic right ventricular cardiomyopathy was recognized. The values of right ventricular EF measured in 3D were 29,1- 60%, mean 44,7 ±10,5%, in 2D: 28,9-71% mean 51 ± 13% and in CMR 25-53% mean 41,9 ± 11,2%. The reduction of EF in 3D from 22,9% to 42% was observed in patients with complex ventricular arrhythmia, while in children with supraventricular arrhythmia or single ventricular beats EF was from 48,5% to 60%. Very high significant correlation ($R = 0.89$, $p < 0.05$) between values of EF obtained by 3D-echocardiography and CMR was found. High significant correlation between 3D EF and Simpson 2D EF ($R = 0.66$, $p < 0.05$) was observed.
Conclusions: 1. Impaired right ventricular function is observed in patients with severe ventricular arrhythmia. 2. 3 D-echocardiographical analysis of right ventricular function may be valuable tool for clinical assessment of children with arrhythmia.

P2418 - THE ASSESSMENT OF CHILDREN WITH HYPERTROPHIC CARDIOMYOPATHY ASSOCIATIONS OF SPECKLE TRACKING ECHOCARDIOGRAPHY AND LATE GADOLINIUM ENHANCEMENT CARDIAC MAGNETIC IMAGING

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Background: We sought to identify surrogate markers of left ventricular (LV) fibrosis in children with hypertrophic cardiomyopathy (HCM) using 2D speckle tracking and to clarify the relationship between deformation indices, extent of LV myocardial fibrosis and hypertrophy.
Methods: Twenty-seven patients under the age of 18 years with an established diagnosis of HCM, who underwent both echocardiographic and CMR investigations, were enrolled in this single tertiary centre retrospective study. Twenty-seven age- and sex-matched healthy controls had an echocardiogram only. Myocardial strain was analysed against the extent of fibrosis assessed by late gadolinium enhancement at MRI and myocardial thickness.
Results: Patients had a significantly higher mean (SD) ejection fraction of 70.8% (8.2) vs 61.2% (6.2), $p < 0.001$ and shortening fraction than controls (44.7%, IQR (37.0, 48.5) vs 36.7% (32.0, 41.7%, $p = 0.001$), but significantly worse global longitudinal (median -18.0, IQR(-14.6,-20.6) vs -21.5 (-19.1,-22.5), $p = 0.002$), radial (28.1, SD(9.4) vs 44.6, SD(11.6); $p < 0.001$) and circumferential strain (-23.8, SD(5.3) vs -27.8, SD(5.6); $p = 0.009$). Fibrosis was present in 11 of 25 patients (44%). The interaction of segmental fibrosis and hypertrophy was associated with attenuated radial ($p = 0.037$) and longitudinal strain ($p = 0.001$). When myocardial segments were analysed, the radial strain diminished significantly as the amount of fibrosis increased ($p = 0.04$). No association was found between the extent of segmental fibrosis, myocardial thickness, and circumferential and longitudinal strain.
Conclusions: Myocardial deformation indices are decreased in patients with HCM. Myocardial fibrosis and hypertrophy have an independent impact on global and regional systolic function however it is likely that they are not the only factors to play a role.

P2430 - CARDIAC EVALUATION FOR DETECTION OF CONGENITAL HEART DISEASE IN ASYMPTOMATIC TERM NEWBORNS A NEW LOOK AT THE ALGORITHM

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Background: The incidence of congenital heart disease (CHD) is 1% of live births in the United States. Of these, 25% are major or critical CHD. In 1994, the American Heart Association produced guidelines for the evaluation of CHD in neonates. A lot has changed since then, including widespread use of fetal echocardiogram and screening pulse oximetry for detection of CHD. We evaluate the value of these tests in today's context and propose a revised algorithm for the detection of CHD.
Methods: We reviewed asymptomatic term newborns who had cardiac evaluation including electrocardiogram (ECG) and echocardiogram, from 2013 to 2016. Murmurs were categorized as benign sounding (innocent) or pathologic based on the intensity and quality of the murmur. ECG findings were divided into normal, QRS abnormality and other abnormalities (Table 1). Echocardiograms were classified into 4 groups – Normal study, Minor, Major or Critical anomaly (Table 2).
Results: 777 newborns underwent cardiology evaluation during the study period. All the patients had echocardiogram and 676 had ECG. Among these, 3 critical, 8 major and 517 minor CHD were identified. The distribution of CHD, correlated with type of murmur and ECG is described in Table 3. All major and 1 critical CHD were present in patients with pathological murmur. Two patients with innocent murmur and abnormal ECG had critical CHD. Patients without any murmur with or without abnormal

Table 1. Categorization of Electrocardiogram findings.

QRS voltage Abnormality (n = 283)	Right ventricular hypertrophy
	Left ventricular hypertrophy
	Septal hypertrophy
	Biventricular hypertrophy
Other abnormalities (n = 79)	Sinus bradycardia or tachycardia
	Non-specific ST-T wave changes, Prolonged QTc
	Axis deviation: right or teft
	Premature atrial contraction, Premature ventricular contraction
	Atrial flutter. Supraventricular tachycardia
Normal (n = 314)	Normal ECG and normalvariants

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ECG, those with innocent murmur and normal ECG did not have any significant CHD.

Conclusion: Pathologic murmurs when present should be evaluated by echocardiogram. ECG may be helpful in detecting critical CHD in newborns with innocent murmur. Newborns with innocent murmur and normal ECG do not have major CHD and can be followed as outpatient. Based on our findings, we propose an algorithm for a systematic approach to cardiac evaluation of asymptomatic term newborns. (Figure 1).

Table 2. Categorization of Congenital Heart Disease

Critical (n = 3)	
Major (n = 8)	any CHD requiring immediate intervention Example: critical aortic or pulmonary stenosis, hypoplastic left heart syndrome, transposition of the great arteries any CHD requiring intervention within 6 months of life Example: large VSD, atrioventricular septal defect tetralogy of fallot
Minor (n = 517)	any CHD expected to resolve spontaneously or unlikely to require intervention Example: small PDA, ASD or small muscular VSD.
Normal (n = 249)	No CHD Example: PPO, very small PDA and normal variants

Table 3. Distribution of Congenital Heart Disease by Type of Murmur and ECG Findings.

	No ECG	**Normal ECG**	**Abnormal ECG (QRS abnomality**	**Abnormal ECG (Other abnormalities**	**Total**
Other reasons for consult (No murmur)	Total = 39 Minor = 24 Major = 0 Critical = 0	Total = 135 Minor = 70 Major = 0 Critical = 0	Total = 132 Minor = 77 Major – 0 Critical = 0	Total 49 Minor = 25 Major = 0 Critical = 0	Total = 355 Minor = 196 Major = 0 Critical = 0
Benign murmur	Total = 45 Minor = 30 Major = 0 Critical = 0	Total = 124 Minor = 91 Major = 0 Critical = 0	Total = 122 Minor = 86 Major = 0 Critical = 2	Total 27 Minor = 21 Major = 0 Critical = 0	Total = 318 Minor = 228 Major = 0 Critical = 2
Pathologic murmur	Total = 17 Minor = 15 Major = 1 Critical = 0	Total = 55 Minor = 51 Major = 4 Critical = 0	Total = 29 Minor = 24 Major = 3 Critical = 1	Total 3 Minor = 3 Major = 0 Critical = 0	Total = 104 Minor = 93 Major = 8 Critical = 1
Total	Total = 101 Minor = 69 Major = 1 Critical = 0	Total = 314 Minor = 212 Major = 4 Critical = 0	Total = 283 Minor = 187 Major = 3 Critical = 3	Total 79 Minor = 49 Major = 0 Critical = 0	Total = 777 Minor = 517 Major = 8 Critical = 3

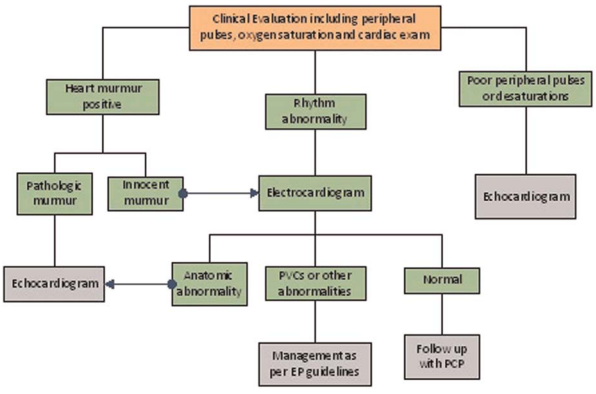

Figure 1.
Algorithm for Evaluation of CHD in Asymptomatic Term Newborn Babies.

P2439 - DEVELOPMENT OF A NOVEL INKJET PRINTING SYSTEM TO REALIZE MASS PRODUCTION OF SUPER FLEXIBLE HEART REPLICAS FOR SURGICAL SIMULATION OF COMPLICATED CONGENITAL HEART DISEASE

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Backgrounds: Precise understanding of 3D anatomy is crucial for successful surgical operation of congenital heart diseases. We have developed a new technology that reproduces extremely flexible polyurethane replicas of congenital heart disease by employing stereolithography 3D printing followed by vacuum casting techniques. However, this technique takes approximately one week of time and costs considerable amount of money. Here we introduce a new inkjet printing system which is exclusively devised for manufacturing super flexible heart replicas for congenital and adult heart diseases.

Methods: In collaboration with Japanese companies SCREEN Holdings Co., Ltd., cross Effect Inc., and Kyoeisha Chemical Co., Ltd., we developed a new ink jet printing machine which consists of 4 print heads being able to manufacture 200mmx500mm super flexible heart replicas for surgical simulation. Four replicas of congenital heart diseases including VSD, sinus venosus ASD, TOF after BT-shunt, and congenitally corrected TGA were experimentally manufactured with this new technique.

Results: The new inkjet printing system allowed to reproducing faithfully these congenital heart diseases within 24 hours, resulting in possible reduction of the manufacturing costs. The texture and elasticity of the replicas were similar to the real heart of children. These heart replicas with ink jet printing enabled surgeons to cut and suture, facilitating the simulation of the surgical operation.

Conclusions: We have successfully devised a new inkjet 3D printing machine as an academia-industry collaboration. Further improvement in 3D printing system, ink, and support materials are necessary to be safely used and approved in the clinical filed.

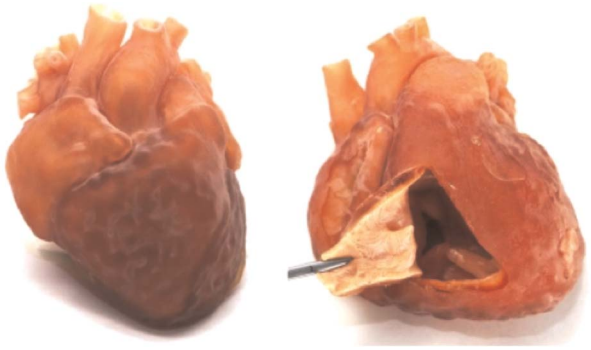

Figure.

P2452 - TRICUSPID VALUE SIZE IN FETUSES WITH PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM SEVERE PULMONARY STENOSIS IS A STRONG PREDICTOR OF POSTNATAL OUTCOME

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Background: Pulmonary atresia with intact ventricular septum (PAIVS) and severe pulmonary stenosis (SPS) are strongly influenced by the prediction of univentricular (UV) or not-UV.
Objective: The purpose of this study was to determine the morphologic and physiological predictors of post-natal surgical pathway, presence or absence of ventriculocoronary connections (VCC) of fetuses with PAIVS and SPS.
Materials: We identified 16 fetuses (gestational age 29.3 weeks), with a fetal diagnosis of PAIVS and SPS from 2002 to 2015.
Methods: Using a z-scores of fetal cardiac measurememts,we calculated tricuspid valve (TV), tricuspid valve/mitral valve(TV/MV) ratio, pulmonary valve/aortic valve (PV/AV) ratio and pulmonary valve, to facilitate prediction of UV or not-UV circulation. We also assessed the presence or absence of VCC.
Result: Two groups; UV included 13 fetuses, not-UV included 3 fetuses. TV ; -3.1 SD ~ -10.8 SD(median -7.7 SD) vs –3.4 SD ~ -2.9 SD (median -3.2 SD)(p = 0.01)(UV vs not-UV). TV/MV ratio; 0.26 ~ 0.63(median0.34) vs 0.55 ~ 0.67 (median0.57) (p = 0.03).PV/AV ratio ; 0.28 ~ 0.90 (median0.43) vs 0.82 ~ 0.98 (median 0.91)(p = 0.01). PV; -11.0 SD ~ 0.27 SD (median-8.7 SD) vs -2.3 SD ~ -1.2 SD (median-1.6 SD)(p = 0.06). A cut-off value of -5SD for TVwas highly predictive of UV circulation, with sensitivity of 92% and a specificity of 100%. A cut-off value of -5.5 SD for TV was highly predictive of VCC during fetal life, with sensitivity of 100% and a specificity of 58%.
Conclusion: Tricuspid valve in fetuses affected by PAIVS andSPS is a strong prenatal echocardiographic predictor of UV circulation and VCC.

P2460 - MORPHOMETRY OF DUCTUS ARTERIOSUS AND ITS ASSOCIATION TO FLOW DYNAMICS A FETAL DOPPLER ECHOCARDIOGRAPHIC CAVEAT

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Background/Hypothesis: The shape of fetal ductus arteriosus is cuneiform, but there is no information correlating flow dynamics to ductal morphometry. This study test the hypotheses that ductal velocities are higher and pulsatility index (PI) is lower at the distal (pulmonary) extremity than at the proximal (aortic) end of the vessel, and that there is correlation between flow dynamics and ductal diameter.
Material and Methods: Normal fetuses from mothers without any comorbities, at any gestational age, were recruited to the study. Difficulty to assess both ductal extremities were cause for exclusion. 2-D echocardiography with color mapping was aimed at determining maximal ductal diameters both at aortic (proximal) and pulmonary (distal) ends of the ductus. Doppler analysis of SV and DV, as well as PI determination, was also performed at both ductal extremities in every case.
Results: The sample was made up of 77 fetuses. Proximal mean ductal diameter was 4.5 ±1.3 mm and distal mean diameter was 2.3 ± 0.7mm, p < 0.001. Proximal mean ductal velocities were : SV = 0.75 ± 0,2 m/s; DV = 0.13 ± 0.04 m/s and distal mean velocities were: SV = 1.03 ± 0.21 m/s; DV = 0.20 ± 0.04 m/s, p < 0.001. Mean PI was 2.70 at the aortic end and 2.30 at the pulmonary end, p < 0.001. Proximal and distal ductal diameters were positively correlated (r = 0.80, p < 0.01). There were inverse correlations between ductal diameter and SV (r = 0.28, p = 0.001) and DV (r = 0.40, p = 0.001). A positive correlation between ductal diameter and flow PI (r = 0.65 p = 0.001) was shown.
Conclusions: This original study demonstrates that, since the ductus is cuneiform in shape, flow impedance is higher (higher velocities and lower pulsatility index) at the distal end, which is the correct place to position the Doppler sample-volume during flow analysis, especially when ductal constriction is sought.

P2463 - LONGITUDINAL SYSTOLIC STRAIN AND DEFORMATION GRADIENT ON 4 CHAMBER 2 D SPECKLE TRACKING IN THALASSEMIA MAJOR

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Background: Myocardial siderosis secondary to recurrent blood transfusions contributes to >70% of the morbidity and mortality in Thalassemia Major (TM) patients. 2D Speckle Tracking Echocardiography (2D STE) may be useful for assessment of preclinical myocardial dysfunction.
Methods: TM patients were recruited for the AMIT study (efficacy of amlodipine with chelation in reducing myocardial iron). All patients underwent standard echocardiographic imaging at baseline, 6 and 12 months. Global and segmental longitudinal systolic strain (LSsys) was measured in the left ventricle (LV) (apical 4 chamber). The ratio between mean apical LSsys and basal LSsys (LSsys api/bas) (medial and lateral wall) was calculated.
Results: A total of 19 TM patients completed the AMIT study. The median age was 16 (8-21) years. 25 data points were available from 2D STE. The mean global LSsys was -20.9% ± 2.2. The medial basal LSsys (-17.3% ± 2.2) and lateral basal LSsys (-16.7% ± 4.4) strain was significantly lower when compared with mean apical LSsys strain (-24.9% ± 4.0) (p <0.01). Deformation gradient in the LV was seen by a lateral LSsys api/bas ratio of 1.5 ±0.3 and medial LSsys api/bas ratio of 1.6 ±05.
Conclusion: 2D STE demonstrated a pattern of systolic septal longitudinal base-to-apex strain gradient with preserved LSsys at apical segments and significantly reduced basal strain in TM patients. This gradient may help in differentiating TM patients who are at risk for increased cardiovascular morbidity and mortality.

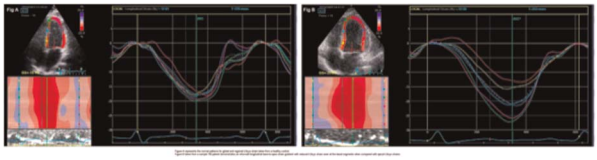

Figure.

P2472 - LOAD DEPENDENT MECHANICAL DYSSYNCHRONY AND MYOCARDIAL DEFORMATION OF LEFT VENTRICLE IN ASYMPTOMATIC ADULT FONTAN PATIENTS WITH TRICUSPID ATRESIA

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Fontan failure as a late complication of Fontan operation represents a significant challenge to manage. We evaluated

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load-dependent mechanical dyssynchrony, myocardial deformation of left ventricle (LV) and the change of Fontan circulation in asymptomatic adults with tricuspid atresia (TA) after Fontan procedure. Comparative analysis was performed between 29 asymptomatic tricuspid atresia (type 1) patients with normal LV ejection fraction (>50%) and 23 normal healthy controls. After baseline measurements, isometric hand grip test (IHGT) was used to increase LV afterload. Subsequently, sublingual nitroglycerin was administered to modify preload. Conventional echocardiographic parameters, LV strain, LV dyssynchrony index and LV torsion, were calculated under each condition. At baseline, in the TA group diastolic sphericity index increased (0.74 ± 0.13 vs. 0.51 ± 0.03; $P < 0.001$)and LV longitudinal strain decreased ($-16.7 \pm 2.3\%$ vs. $-20.0 \pm 2.0\%$; $P < 0.001$). The difference in time-to-peak radial strain between the anteroseptal and posterior segments (LVdys-2) (32.4 ± 73.4 ms vs. 3.5 ± 9.9 ms; $P = 0.045$) and the absolute difference in time-to-peak radial strain for 6 segments (LVdys-6) (62.3 ± 88.9 ms vs. 8.2 ± 15.0 ms; $P = 0.003$) increased in the TA groups compared to the control group. At baseline, LV torsion ($12.6 \pm 7.9°$ vs. $19.2 \pm 8.4°$; $P = 0.006$), basal rotation ($-1.1 \pm 2.4°$ vs. $-4.1 \pm 2.6°$; $P = 0.0001$), twist rate ($50.9 \pm 27.0°/s$ vs. $84.5 \pm 30.3°/s$; $P < 0.0001$), untwist rate ($51.2 \pm 28.7°/s$ vs. $73.9 \pm 30.1°/s$; $P = 0.008$), and torsional rate ($76.0 \pm 36.9°/s$ vs. $124.0 \pm 37.9°/s$; $P < 0.0001$) decreased in the TA patients. The repetitive measured-ANOVA by Greenhouse-Geisser method among baseline, IHGT, and nitroglycerin administration showed the different load-dependent responses between two groups about LV longitudinal strain (group $P < 0.001$,within $P = 0.848$, interaction $P = 0.045$), LVdys-6 (group $P = 0.042$, within $P = 0.026$, interaction $P = 0.379$), and LV torsion (group $P = 0.027$ within $P = 0.106$, interaction $P = 0.058$). In conclusion, although asymptomatic adult TA patients after Fontan procedure had normal LV ejection fraction, their LV deformations were impaired and abnormal load-dependent responses were also demonstrated.

P2476 - NORMAL SEGMENTAL MYOCARDIAL DEFORMATION IS NOTED IN PATIENTS WITH ANOMALOIS AORTIC ORIGIN OF CORONARY ARTERIES DESPITE MALIGNANT COURSE AND ISCHEMIC SYMPTOMS

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Background: Sudden cardiac death is the first presenting symptom in few with Anomalous aortic origin of coronary artery (AAOCA), Most being incidentally diagnosed for non-ischemic complaints. We hypothesized segmental longitudinal (SLS) and circumferential strain (SCS) abnormalities would be present at baseline in AAOCA patients with past ischemic events.
Method: Two-dimensional speckle tracking (2DSTE) derived strain assessment was performed using GE Echopac software on 16 left ventricular segments from apical and 6 mid wall segments from the short axis views. Less than 10% shortening for SLS and SCS was considered abnormal. Descriptive analysis of segmental strain abnormalities during rest, correlation with the presence of baseline symptoms, exercise stress test and finding on CT angiogram were performed.
Results: 25 Pts. with AAOCA. 20(80%) males, 5(20%) females. Age range 7-19. 9(36%)pts. were symptomatic. 20(80%)Pts. with AAO of Right coronary (AAORCA), 3(12%)Pts. had AAOLCA, 2(8%)Pts. had both. 1(4%)Pts. had abnormal Exercise stress test. 5 (20%)Pts. had significant coronary stenosis (>50%) on CT angiogram. 3(12%)Pts. needed surgical intervention. No abnormalities in SLS were noted by 2DSTE in those with AAORCA. Only 3 (15%)Pts. had SCS abnormalities (figure). Out of three patients with abnormal findings only one presented with ischemic symptom's and another one had >50% luminal narrowing. The basal lateral and inferolateral segment abnormalities in the circumflex territory were noted in 85% patients with AAORCA.
Conclusion: Normal SLA and SCS was noted in the anomalous coronary territory in patients with AAORCA despite presence of symptoms or evidence of >50% stenosis on CTA. False positive results are noted by this technique in areas not supplied by the anomalous coronary. Two patients with AAOLCA did have ischemic symptoms and abnormal segmental findings. Further studies are recommended in a larger cohort to assess the prognostic value of segmental deformation in patients with AAOLCA.

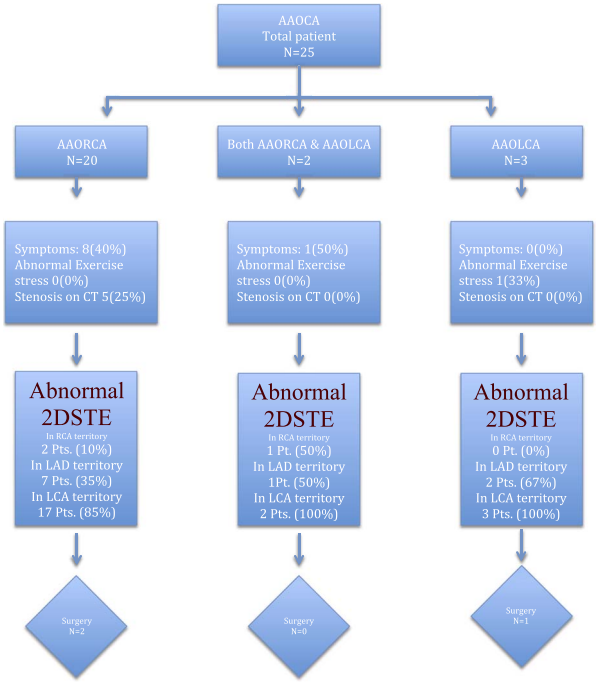

Figure.

P2482 - CLINICALLY RELEVANT BRAIN INJURY AND DEVELOPMENTAL ABNORMALITIES ARE COMMON IN AN UNSELECTED CLINICAL COHORT OF NEONATES WITH CONGENITAL HEART DISEASE

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Background: Neonates with CHD studied with MRI in the neonatal period are recognized to be at high risk of brain injuries pre- and post-operatively, reported in 30% to 50%. However, the frequency of brain abnormalities in unselected clinical cohorts of neonates with CHD remains unknown. These data are needed to

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appropriately evaluate the use of brain MRI in routine clinical practice for neonates with CHD requiring cardiac surgery.
Objective: To determine the rate of injury and developmental anomalies on clinical pre- and post-operative MRI obtained routinely in a consecutive series of term neonates with CHD.
Design/Methods: Since March 2015, brain MRI has been introduced as the standard of care in a large tertiary pediatric hospital. Neonates undergoing cardiac surgery in the first few weeks of life undergo an MRI before and after the surgical intervention as soon as they are stable enough for transportation. Brain abnormalities, such as white matter lesions (WMI), intraventricular hemorrhage (IVH), arterial stroke, sinus venous thrombosis (SVT), hydrocephalus, hematomas, and developmental anomalies are recorded systematically. Key demographic and clinical data are collected from the clinical charts, independent of the MRI reviews.
Results: 106 neonates underwent clinical brain MRI: median gestational age 39 weeks, birth weight 3.3 kg, and 67 males. The types of CHD were: 39 transposition of the great arteries (TGA), 28 single ventricle physiology (SVP) and 39 other cardiac lesions. Preoperative brain injury was common: WMI in 8 (2 TGA, 2 SVP, 4 others), IVH in 3 (1 TGA, 1 SVP, 1 others), stroke in 6 neonates (2 TGA, 2 SVP, 2 others). Sinus venous thrombosis was seen in 1 neonates with SVP. Subdural hematoma was diagnosed in 7 (2 TGA, 2 SVP, 3 others). Brain anomalies consistent with tubulinopathy were seen in 1 patient. New post-operative injuries were similarly frequent: WMI in 4 children (3 TGA,

P2485 - HIGH INCIDENCE OF ABNORMAL TEI INDEX IN THE FETUSES WITH MATERNAL ANTI SS A ANTIBODY

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Maternal anti SS-A antibody affects to the fetal heart and causes heart block and myocarditis. Detection of the damage to the atrioventricular node is well documented. However, the incidence of the myocardial damage is not well known. Therefore, we investigated the cardiac function in the fetuses with maternal anti SS-A antibody.
Method: This is a retrospective study in single center. We reviewed our clinical record of fetal echocardiography in 5 years from Jan. 2012 to Dec 2016. In this period, 24 cases had fetal echocardiography due to maternal positive anti SS-A antibody without fetal heart block. Of the 24, 18 cases had sequential examination from second trimester. Tei index of the right and left ventricle were measured. Postnatal data were available in 20 cases.
Results: Of the 24, first examination were 18w3d to 38w1d (median 22w0d). During pregnancy, 2 cases (8%) had high Tei index >0.7, and other 10 (42%) had relatively high Tei index >0.6. Of these 12, 6 had Tei index >0.6 also at the last examination before birth, but, the other had transient elevation. 20 with the postnatal data were born at 36w5d to 41w2d (median 39w2d), and their birth weight were 1918g to 3336 g (mean 2778 g). No case had clinical symptom of cardiac dysfunction, although one had transient tachypnea, one had small for gestational age, and the other had premature rupture of membrane.
Conclusions: Our data suggest high incidence of abnormal Tei index in the fetuses with maternal anti SS-A antibody. Although the cardiac dysfunction seems to be transient and subclinical in most of the cases, further study with more detail fetal cardiac functional study and postnatal follow-up are required.

P2495 - QUANTIFICATION OF RIGHT VENTRICULAR ELECTRO MECHANICAL DYSSYNCHRONY AND ITS RELATION TO CLINICAL OUTCOMES IN CHILDREN WITH REPAIRED TETRALOGY OF FALLOT

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Introduction: Electro-mechanical dyssynchrony occurs in patients following tetralogy of Fallot (TOF). The rightward septal flash, with early septal activation and post-systolic shortening of the right ventricular (RV) basal lateral wall, is ubiquitous in this population and contributes to inefficient RV mechanics. The aim of the study was to quantify the severity of electro-mechanical dyssynchrony in relation to RV remodelling, dysfunction and clinical outcomes in a pediatric cohort following TOF repair.
Methods and Results: A retrospective review of prospectively collected data was performed in 81 children post-TOF repair, aged 13.6 ± 2.9 years, and compared to 50 pediatric controls. Two-dimensional echocardiography speckle-tracking, cardiac magnetic resonance (CMR) and clinical details were obtained. Patients had higher pre-stretch and post-systolic strain (PSS) amplitude and duration, mechanical dispersion and basal lateral-septal wall time-to-peak strain differences than controls (p-values all <0.001). All intra-RV dyssynchrony timing parameters were associated with reduced RV ejection fraction (EF) by CMR and/or global RV longitudinal strain. The pre-stretch duration as a percentage of total shortening time and the time-to-peak strain differences between the basal lateral RV and mid-septal segments were independently associated with RV EF using multivariate analysis. PSS amplitude was higher in patients with arrhythmias compared to arrhythmia-free patients (7.8(4.2-13)% vs 2.0(0-12.5)%, $p = 0.03$). Intraventricular dyssynchrony parameters were not associated with myocardial fibrosis by late gadolinium enhancement on CMR.
Discussion: The right-sided septal flash is viewed as a dichotomous variable, however we sought to describe a continuum of dyssynchrony severity and its relationship to pathophysiology and clinical outcomes in repaired TOF patients. RV basal lateral-septal timing delays, pre-stretch and PSS may provide a practical approach to assessing RV electro-mechanical dyssynchrony severity and one which reflects the underlying pathophysiology.

P2511 - ECHOCARDIOGRAPHIC PREDICTION OF USE TRANSANNULAR PATCH DURING TETRALOGY OF FALLOT REPAIR

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Background: The long-term outcome after repair of tetralogy of Fallot (TOF) is critically dependent on the competence of the pulmonary valve that is compromised by trans-annular patch (TAP).
Objectives: To determine the echocardiographic predictors of TAP in infants undergoing TOF repair.
Methods: Consecutive infants operated for TOF were prospectively included. Aortic and pulmonary annuli (PAnn), and

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main pulmonary artery (MPA) were measured preoperatively and their z scores were determined from published nomograms. A novel index, the pulmonary annulus index (PAI) was calculated by dividing observed PAnn from expected PAnn [(Aortic + PA annulus) /2]. The decision to place a TAP was based on intraoperative sizing with Hegar dilators with surgeons blinded to PAI values. Aggressive valve preservation was attempted in borderline situations. Receiver operator curves (ROC) were generated for all three indices.
Results: Of 69 infants (8 ± 2 months), 32 needed TAP (46%). In-hospital mortality was 2/32 in the TAP group and 1/39 in the rest. The performance of individual indices is shown (table). Of 27 infants with PAI ≥ 0.8, three needed a TAP. Their MPA z scores were -5.8, -6 and -6. A PAI < 0.8 was however not consistently associated with requirement of TAP. Of the 42 with PAI <0.8, TAP could still be avoided in 13 patients. The performance of both PAI and PAnn Z scores (cut off: -1.67) were quite similar in predicting the need for TAP (table).
Conclusions: A pulmonary annulus index of ≥ 0.8 can be used to predict the avoidance of TAP in infants undergoing TOF repair especially when combined with MPA z scores. However a low PAI (<0.8) does not necessarily mandate the requirement of a TAP.

Table. Performance of individual echo parameters in predicting need for Trans-Annular Patch.

Variable	Sensitivity (%)	Specificity (%)	Positive predictive value (%)	Negative predictive value (%)	Odds ratio with 95% CI	Area under the curve
PAnn z <-1.67	87	64.9	68.3	85.7	12.9 (3.7-44.9)	0.79
PAI <0.8	87.5	65	68	85.7	11.3 (4 – 47)	0.80
MPA z <-3.0	72	65	64	72	4.7 (1.7-13.1)	0.81

PAnn: Pulmonary Annulus; PAI: Pulmoary Annulus Index, MPA: Main Pulmonary Artery, CI: Confidence Intervals.

P2513 - EXPERIMENTAL PULMONARY INSUFFICIENCY LEADS TO LEFT VENTRICULAR COMPRESSION AND IMPAIRED RELAXATION

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Background: In repaired tetralogy of Fallot with pulmonary insufficiency (PI) left ventricular (LV) dysfunction is a recognized risk factor for exercise intolerance, morbidity, and sudden death. However, the role of PI in the development of LV dysfunction is incompletely understood. The aim of this study is to assess serial changes in LV function and interventricular interactions in an experimental rat model of isolated severe PI.
Methods: Severe PI, defined by echo-Doppler color flow reversal starting in the branch pulmonary arteries, was induced in 6-week-old male Sprague-Dawley rats by puncturing the pulmonary valve leaflets with a 22.5 G needle inserted through main pulmonary artery. Rats underwent echocardiography before, and every 2 weeks after the procedure for 12 weeks (Vivid-E9 (GE)) using a 12-MHz phased array transducer. RV and LV end-diastolic area (RVEDa/LVEDa) were measured in a short axis view and corrected for BSA. Both ventricular systolic and diastolic event timing corrected for cardiac cycle length (CCL) and Tei-index were analyzed using spectral Doppler. LV ejection fraction (LVEF), peak LV free-wall s' and e' velocities, mitral inflow E and A wave peak Doppler velocities were measured and the E/A and E/e' ratios were calculated. LV global longitudinal (LVGLS) and circumferential (LVGCS) strain were measured using 2-D speckle tracking. Results were compared with sham controls using two-way repeated measures ANOVA.
Results: In PI rats (n = 5), RVEDa was larger, while LVEDa was smaller vs. shams (n = 5), and there was a negative correlation between RVEDa and LVEDa (r = -0.41, P = 0.014). Isovolumic relaxation time (IRT) was longer in PI rats vs. shams . As a result, the LV-Tei index was higher in PI rats vs. shams. Other parameters were similar between the groups (Table 1).
Conclusions: Experimental isolated severe PI induces RV dilatation, which compresses the LV and leads to impaired LV relaxation.

Table 1. Results of serial cchocardiographic measurements

	Baseline		2 weeks		4 weeks		6 weeks	
	sham	PI	sham	PI	sham	PI	sham	PI
LVEDA (cm2/m2)	10.8 ± 1.9	9.9 ± 1.2	11.0 ± 1.6	9.0 ± 1.0	10.4 ± 0.8	9.5 ± 0.8	10.6 ± 0.9	9.6 ± 0.9
RVEDA (cm2/m2)	3.8 ± 0.6	4.3 ± 1.5	3.6 ± 1.3	8.5 ± 2.5	3.7 ± 0.8	9.7 ± 0.6	3.9 ± 0.7	8.5 ± 0.5
LVEF (%)	61 ± 2	69 ± 4	72 ± 7	71 ± 6	66 ± 5	71 ± 9	63 ± 3	66 ± 8
s' (mm/s)	45 ± 8	40 ± 10	52 ± 8	60 ± 16	67 ± 24	62 ± 19	61 ± 10	61 ± 12
E (mm/s)	102 ± 2	77 ± 3	103 ± 10	101 ± 18	112 ± 8	104 ± 11	110 ± 20	97 ± 39
A (mm/s)	90 ± 28	99 ± 31	88 ± 20	74 ± 13	90 ± 31	78 ± 11	92 ± 28	94 ± 19
E/A	1.5 ± 0.4	1.2 ± 0.6	1.3 ± 0.1	1.5 ± 0.5	1.7 ± 0.3	1.4 ± 0.3	1.2 ± 0.4	1.1 ± 0.4
e' (mm/s)	NA	NA	45 ± 3	50 ± 16	43 ± 2	51 ± 18	42 ± 14	37 ± 7
E/e'	NA	NA	23 ± 1	24 ± 10	26 ± 2	24 ± 13	26 ± 7	26 ± 10
LVGLS (%)	17.9 ± 2.3	15.9 ± 1.1	19.5 ± 1.2	19.4 ± 6.0	18.7 ± 5.2	18.4 ± 0.7	18.5 ± 0.8	20.9 ± 0.9
LVGCS(%)	17.4 ± 1.5	19.2 ± 3.3	17.2 ± 1.9	18.5 ± 4.5	18.0 ± 2.9	17.8 ± 1.4	16.4 ± 3.4	17.8 ± 2.4
LV ET (%)	46 ± 1	50 ± 4	44 ± 5	43 ± 5	45 ± 3	43 ± 2	43 ± 4	43 ± 4
LV FT (%)	42 ± 4	41 ± 4	44 ± 3	47 ± 4	47 ± 5	46 ± 4	48 ± 5	45 ± 6
LV IRT (%)	4.2 ± 4.4	6.0 ± 1.4	3.4 ± 3.9	5.5 ± 3.3	4.9 ± 4.1	5.0 ± 3.0	3.2 ± 3.8	6.5 ± 4.4
LV ICT (%)	6.4 ± 1.5	0.4 ± 5.3	5.6 ± 2.2	4.6 ± 2.6	2.4 ± 3.4	6.4 ± 2.9	4.0 ± 2.9	4.6 ± 1.4
LV Tei index	0.23 ± 0.09	0.12 ± 0.09	0.21 ± 0.11	0.24 ± 0.03	0.16 ± 0.05	0.28 ± 0.10	0.16 ± 0.06	0.26 ± 0.11

	8 weeks		10 weeks		12 weeks		mean		
	sham	PI	sham	PI	sham	PI	sham	PI	*P*-value
LVEDA (cm2/m2)	10.2 ± 1.1	9.0 ± 1.1	10.1 ± 0.7	8.7 ± 1.1	9.4 ± 0.5	8.6 ± 0.5	10.4 ± 1.1	9.2 ± 1.0	< 0.001
RVEDA (cm2/m2)	3.3 ± 0.6	7.8 ± 2.2	3.6 ± 1.0	8.5 ± 3.0	4.1 ± 0.5	8.6 ± 1.7	3.7 ± 0.8	8.0 ± 2.4	< 0.001
LVEF (%)	67 ± 5	65 ± 3	66 ± 2	74 ± 6	70 ± 6	72 ± 8	67 ± 5	69 ± 7	0.139
s' (mm/s)	62 ± 6	75 ± 29	51 ± 9	71 ± 13	55 ± 14	67 ± 22	57 ± 13	63 ± 19	0.124
E (mm/s)	103 ± 3	101 ± 14	84 ± 7	95 ± 9	78 ± 17	106 ± 19	100 ± 15	99 ± 19	0.852
A (mm/s)	90 ± 17	90 ± 27	79 ± 24	97 ± 24	103 ± 39	92 ± 9	90 ± 26	89 ± 21	0.852
E/A	1.2 ± 0.3	1.3 ± 0.2	1.3 ± 0.3	1.2 ± 0.2	1.4 ± 0.7	1.1 ± 0.1	1.3 ± 0.3	1.3 ± 0.3	0.390
e' (mm/s)	42 ± 4	43 ± 10	38 ± 8	55 ± 18	41 ± 16	50 ± 20	42 ± 8	47 ± 15	0.186
E/e'	25 ± 3	26 ± 3	23 ± 4	21 ± 4	25 ± 6	26 ± 14	24 ± 4	25 ± 9	0.895
LVGLS (%)	20.3 ± 1.4	19.8 ± 0.9	18.3 ± 1.3	17.4 ± 0.2	20.2 ± 1.1	18.4 ± 0.03	19.0 ± 2.2	18.6 ± 0.3	0.588
LVGCS(%)	18.1 ± 3.4	17.3 ± 1.4	16.1 ± 1.2	18.2 ± 0.3	17.5 ± 2.3	18.7 ± 0.3	17.2 ± 2.2	18.2 ± 2.1	0.276
LV ET (%)	41 ± 4	43 ± 4	43 ± 5	43 ± 4	47 ± 6	42 ± 2	44 ± 4	44 ± 4	0.634
LV FT (%)	47 ± 4	46 ± 5	48 ± 5	44 ± 4	46 ± 3	46 ± 3	46 ± 4	45 ± 4	0.252
LV IRT (%)	5.0 ± 1.5	9.0 ± 2.7	4.4 ± 1.0	7.1 ± 0.4	4.2 ± 1.8	7.4 ± 1.9	4.2 ± 2.9	6.6 ± 2.7	0.002
LV ICT (%)	6.0 ± 0.9	3.3 ± 2.6	4.4 ± 2.4	4.5 ± 3.4	4.2 ± 1.6	3.8 ± 1.7	4.7 ± 2.5	3.9 ± 3.2	0.290
LV Tei index	0.26 ± 0.01	0.30 ± 0.13	0.20 ± 0.05	0.27 ± 0.08	0.19 ± 0.08	0.27 ± 0.05	0.20 ± 0.07	0.25 ± 0.10	0.020

Abbreviation; LV: keft venricular, RV: right ventricular, EDA: end-diastolic area, EF: ejaction fraction, GLS: global circumferential strain, ET: ejection time, FT: filling time, IRT: isovolumic relaxation time, ICT: isovolumic contraction time.

P2516 - CHANGE OF MYOCARDIAL DEFORMATION AS AN EARLY MARKER OF MYOCARDIAL DYSFUNCTION IN HYPERTROPHIC CARDIOMYOPATHY IN CHILDREN

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Background: currently, according to the recommendations of the ESC Guidelines, the treatment of hypertrophic cardiomyopathy is exclusively symptomatic. Symptomatic treatment of the disease is not optimal for the patient. It is necessary to prevent his disease progression, but not to «go on about the symptoms». This requires to identify the earliest markers of myocardial systolic dysfunction that will assist in determining the indications for an early start of treatment.

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Materials and Methods: 38 children with hypertrophic cardiomyopathy were examined. The mediana of age was 9 years. We studied left ventricular segmental radial deformation and thickness of myocardial segments. The greatest clinical and diagnostic interest is the thickness of the myocardium segment, in which happened the most rapid (critical) decrease in radial strain values. For this derivative was estimated regression on the data and find the minimum point of the derivative function of myocardial deformation depending on its thickness.
Results: we present a new methodological approach to the examination of children with hypertrophic cardiomyopathy, aimed at early detection of local myocardial systolic dysfunction. For its base taken the study of radial myocardial segmental strain, which is most informative reflects the change in systolic function of the hypertrophic heart wall. The number of patients (N = 38) is currently insufficient to make reasoned opinion about the obtained values of the critical point (at the moment it is found to be Z = 4.24). However, the number of observations is enough to talk about the possibility of applying the method and its use for the future perspectives to obtain the critical value of the myocardium segment thickness in which firstly appears systolic segmental myocardial dysfunction in children with hypertrophic cardiomyopathy.

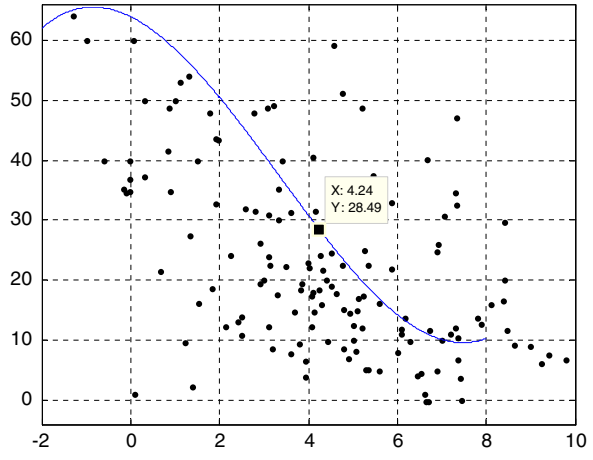

Figure 1.
The radial deformation in depending on the myocardial segment thickness (with the critical point of the myocardium segment thickness, in which happened the most rapid (critical) decrease in radial strain values). Vertical axis – radial segmental strain in %; horizontal axis –thickness of myocardial segment in Z-score).

P2520 - IS THREE DIMENSIONAL TRANSESOPHAGEAL ECHOCARDIOGRAPHY NECESSARY IN TRANSCATHETER DEVICE CLOSURE OF MULTIPLE ATRIAL SEPTAL DEFECTS

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Background: Real time three-dimensional transesophageal echocardiography (RT 3D TEE) allows for evaluation of various shapes of atrial septal defect (ASD) especially in patients with multiple ASDs. During procedure of transcatheter device closure, intracardiac echocardiography (ICE) and/or TEE are integral parts of the assessment of patients with ASD undergoing intervention. In this study, the feasibility, efficacy of RT 3D TEE for transcatheter device closure of multiple ASD was evaluated retrospectively.
Methods: Between January 2005 and March 2016, transcatheter ASD closure with device was attempted in 1169 patients in single center institution. Patients who underwent transcatheter device closure as diagnosed multiple ASD were enrolled retrospectively.
Results: Transcatheter device implantations for multiple ASDs were required in 177(15.1%) of total 1169 cases. Among 177 patients, 41(23.1%) patients had more than three defects. 99(55.9%) patients with multiple ASDs were those under 18 years of ages. Of total 177, only 28 patients (15.8%) had the guidance of RT 3 D TEE during transcatheter closure, and for the others ICE was applied for the guidance of transcatheter closure for even multiple ASD. During procedure using RT 3D TEE, the analysis of ASDs successfully demonstrated rims but was limited for visualized whole defect even real time. Full-volume mode of RT 3D TEE had the best views of large/ multiple defects and surrounding anatomy. However, it was limited for postero-inferior defect by artifact around IVC. For applying RT 3D TEE in children, relatively big size of probe tip had the restriction. Under the guidance of ICE without 3D TEE, there was no adverse effect and also residual defects remained insignificantly.
Conclusion: For transcatheter closure of multiple ASD with device, RT 3D TEE provides important information for evaluation of complexed ASD. However,its application during procedure is relatively limited for successful closure.

P2526 - PRENATAL DETECTION AND EARLY OUTCOME OF SERIOUS AND CRITICAL CONGENITAL HEART DISEASE IN HAWAII FROM 2012 2016

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Background: Prenatal diagnosis of congenital heart disease (CHD) improves neonatal outcomes. In Hawaii, the benefit of prenatal diagnosis is greatly enhanced because neonatal heart surgery is not available. If critical CHD is detected in a fetus, it is recommended that the mother be transferred to a cardiac surgical center on the mainland prenatally.
Methods: We reviewed databases for the total number of initial (per pregnancy) antenatal screening ultrasounds and initial fetal echocardiograms done June 2012 through May 2016 at Kaiser Permanente Hawaii (KPHI). We reviewed the fetal and transthoracic echocardiography databases for diagnoses for which intervention is necessary in the neonatal period (critical CHD) or in the first six months (serious CHD). Fetal and postnatal outcomes of all identified patients were documented.
Results: From 6/2012-5/2016, 11873 initial obstetric screening ultrasounds and 583 initial fetal echocardiograms were performed in the KPHI system. The prenatal detection rate of 31 patients (serious + critical CHD) requiring intervention by 6 months of age was 77%. The diagnoses missed prenatally by obstetric screening included 4 moderate-large ventricular septal defects (VSD), 2 coarctation/VSD, and one simple form of tetralogy of Fallot. None were missed by fetal echocardiography. Thus 1 serious/critical CHD diagnosis was missed per 1696 obstetric screening ultrasounds. Of 20

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critical CHD diagnoses requiring neonatal intervention, 18 (90%) were diagnosed by prenatal screening. There were 9 elective terminations and 2 in utero fetal deaths. The remaining 9 patients underwent neonatal surgery, 5 were delivered at the surgical center on the mainland and 4 needed postnatal transport from Hawaii to the mainland. All patients who had surgery are currently alive and well.
Conclusions: Prenatal screening for critical and serious CHD at KPHI in the last 4 years has been successful with higher detection rates than previously reported in the US and internationally.

P2528 - CONTRAST ENHANCED CAROTID ULTRASONOGRAPHY FOR EVALUATION OF VASA VASORUM IN CHILDREN WITH LARGE CORONARY ANEURYSMS DUE TO KAWASAKI DISEASE

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Background: The vasculitis in patients with Kawasaki disease (KD) is suspected to involve the vasa vasorum (VV). The VV network provides a critical route for inflammatory cell infiltration. We recently performed carotid contrast-enhanced ultrasonography (CEUS) in patients with KD and concurrent coronary artery lesions to compare the heterogeneity of the VV network proliferation between patients with KD and controls.
Case: We evaluated a 2-year-old boy with a large left anterior descending coronary aneurysm (z-score: 9.15) and large right coronary aneurysm (z-score: 9.55) due to KD. He had been diagnosed with KD at 19 months of age. In the acute phase, he received intravenous immunoglobulin (2 g/kg/dose), aspirin (30 mg/kg/day), and prednisolone (2 mg/kg/day) on the third day of illness. At the time of this writing, he was receiving anticoagulants (warfarin, aspirin, and clopidogrel) to prevent coronary thrombosis and had experienced no major adverse cardiovascular events. Six months after the acute onset of KD, he exhibited a greater mean carotid intima-media thickness (0.55 mm) than that in age-matched controls. CEUS using perflubutane (Sonazoid; GE Healthcare, Oslo, Norway) enabled real-time visualization and semi-quantitative evaluation of the carotid VV by setting the regions of interest in the lumen (L) and wall (W) of the common carotid artery. These values were used to calculate the ratio (EIW/EIL) of enhanced intensity (EI) (EI = peak intensity − baseline intensity) derived from a time-intensity curve. He developed no side effects of perflubutane administration.
Conclusions: Carotid CEUS may be useful for noninvasive assessment of the proliferation of neovascularization. Such neovascularization suggests active vascular remodeling of the coronary arteries in patients with KD, even small children. Future investigation of the prognostic value of CEUS is warranted.

P2530 - MORPHOANATOMY OF COMMON ARTERIAL TRUNK – A COMPUTED TOMOGRAPHIC STUDY

Saurabh Kumar Gupta[1], Abhinav Aggarwal[1], Gurpreet Singh Gulati[2], Rajnish Juneja[1], S Ramakrishnan[1], Shyam S Kothari[1], Anita Saxena[1], Sanjiv Sharma[2], Sachin Talwar[3], Balram Airan[3]
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Background: Common arterial trunk is classified based on the variation in the origin of pulmonary arteries (PA). Recent reports have brought out difficulties in following these standard classifications. Sinusal origin of PAs as well as crossed PAs is increasingly being recognized. Nevertheless, there is paucity of data from living hearts so as to realize the potential benefits of this knowledge in improving the surgical outcomes of patients with common arterial trunk.
Materials & Methods: In this prospective observational study, 21 patients with common arterial trunk have been recruited so far. Following detailed clinical and echocardiographic examination all these patients underwent computed tomography. All the morphoanatomic details of various components of the anomaly were studied with an emphasis on the origin of coronary and PA with reference to the circumference of the truncal valve as well as the sinotubular junction (STJ).
Results: Fourteen (66%) patients were classified as Collette-Edwards type I while remaining 7 (34%) were Type 2. Truncal valve was tricuspid in 13 (62%), quadricuspid in 6 (28%) and bicuspid in 2 (10%). Majority of patients had left sided aortic arch while it was right sided in 6 (28%). In one patient, right sided arch was interrupted. Sinusal origin of PAs was seen in 11 of 14 (78%) patients with type I. Right PA was seen arising from truncal sinus in 2 (28%) patients with type II. Three (14%) patients had crossed PAs. Two patients of type I had origin of left coronary artery within 2 mm of the PA orifice.
Conclusion: The anatomy of pulmonary and coronary arteries is highly variable in patients with common arterial trunk. Sinusal origin of PAs, seen in high number of patients, may allow direct reimplantation of PA on to the right ventricular outflow tract, possibly without a conduit.

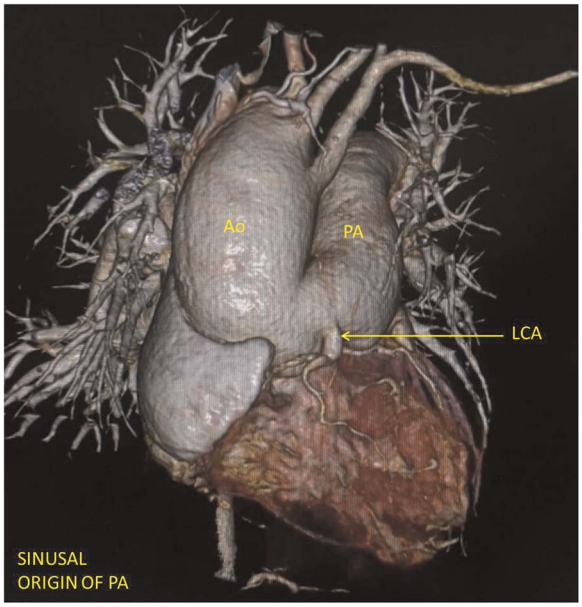

Figure.

P2535 - LARGE LEFT VENTRICULAR MYXOMA DIAGNOSED BY ECHOCARDIOGRAPHY NAWAL A. M. ALABDULKARIM ABDULRAOOF ALSAEEDI MPRINCE SULTAN CARDIAC CENTER RIYADH SAUDI ARABIA

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Clinical Presentation: 9 years old girl presented with sudden hemiparesis due to stroke without previous health problems.
Imaging Findings: Transthoracic Echocardiogram was performed to role out cardiac source of emboli, It showed huge mass filling the whole LV and protruding through aortic valve, it was not homogeneous suggestive of a myxoma.
Role of Imaging in Patient Care: Utilization of echocardiography allowed rapid accurate diagnosis and surgical management then assessment of surgery result via intraoperative transesophageal echocariography and monitoring recurrence at follow up.
Summary and Discussion Points: -This is a case of severe LV outflow tract obstruction due to a LV myxoma originating from the apex. -LV myxoma is extremely rare, accounting for 2.5-4% of all cases. - Left ventricular myxomas present with left ventricular outflow tract obstruction in 55%, embolic phenomena in 55%, and other systemic manifestations in 10%.
Transthoracic parasternal long axis images preoperatively: -The shape, the extension, the site of attachment, the involvement of valve leaflets and the functional obstruction to LV outflow tract could promptly and easily be assessed by echocardiography. -This myxoma required urgent transaortic resection. -The patient was at an increased risk of sudden death and systemic embolization due to the systolic prolapse of the mass through the aortic valve with severe obstruction, the characteristics of the myxoma, and the high pressure within the left ventricle during systole. -The mass was removed by using the transaortic approach in order to avoid a left ventriculotomy and its potential complications. This approach allowed the complete resection of the LV myxoma.

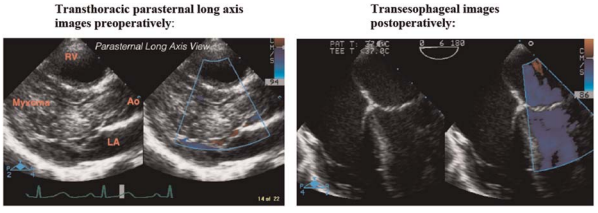

Figure.

P2543 - LARGE NEONATAL PULMONARY ARTERY THROMBUS DIAGNOSED BY ECHOCARIOGRAPHY

Nawal Alabdulkarim
PSCC, Pediatric Cardiology, Riyadh-Saudi Arabia

Clinical Presentation: Full term baby girl delivered to G8P6 + 1 mom by caesarean section due to failure to progress in labour. Baby noted post delivery to have desaturation and heart murmur. Echocardiography confirmed Huge mass seen at distal main pulmonary artery protruding into branch pulmonary arteries causing pulmonary stenosis. No other masses neither abnormalities noted.
Summary: -Pulmonary artery thrombosis in neonates is a rare entity. -Very few cases of spontaneous neonatal pulmonary arterial thrombosis have ever been described. -(The most frequent important risk factor for the development of thrombosis in the neonate is the presence of an indwelling central line. The other important risk factors are asphyxia, septicemia, dehydration, maternal diabetes, and inherited thrombophilias). -When assessing a cyanotic neonate with a structurally normal heart echocardiographically, it is important to look for pulmonary artery thrombi before labeling the study as PPHN. -Most cases of spontaneuous Pulmonary artery thrombosis as in this case located at the bifurcation of the main pulmonary artery in the region of a closed ductus arteriosus with subsequent distal thromboemboli that may have contributed to cyanosis in this neonate. - We expect that the sensitivity for echocardiographic detection of main pulmonary arterythrombi in newborns is higher, as this region is routinely imaged well on newborn echocardiograms,unlike the umbilical vein and ductus venosus.

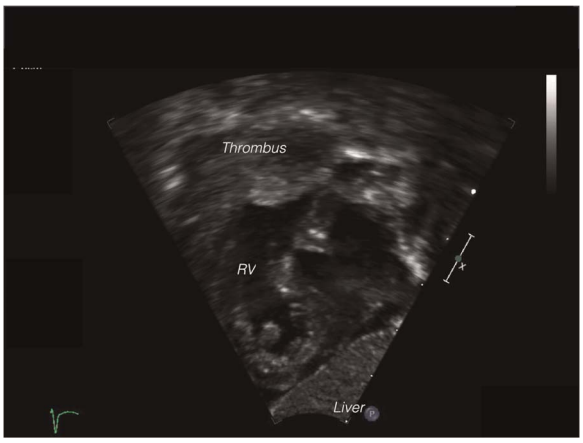

Figure 1.

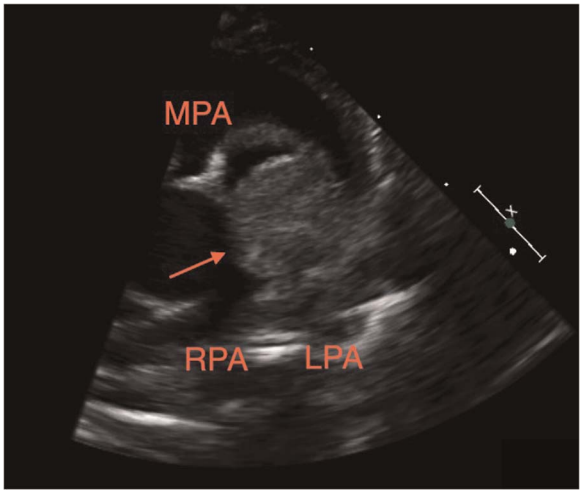

Figure 2.

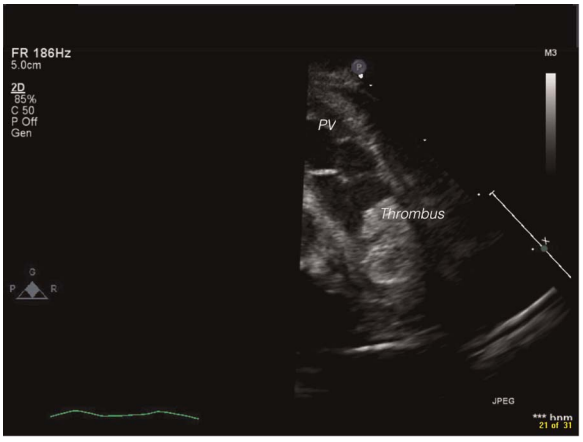

Figure 3.

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P2551 - REDUCTION IN CORONARY BLOOD FLOW IN HOSPITALIZED ADOLESCENTS WITH ANOREXIA NERVOSA COMPARED TO HEALTHY CONTROLS

Liat Gelernter Yaniv[1], Dawod Sharif[2], Jenny Garkaby[3], Camilia Shahla[4], Ameen Khalil[4], Nogah Kerem[1]
Bnai Zion Medical Center, Rappaport Faculty of Medicine, Technion, Pediatrics, Haifa-Israel[1]; Bnai Zion Medical Center, Rappaport Faculty of Medicine, Technion, Cardiology, Haifa-Israel[2]; Bnai Zion Medical Center, Pediatrics, Haifa-Israel[3] Bnai Zion Medical Center, Cardiology, Haifa-Israel[4]

Anorexia nervosa (AN) is an eating disorder characterized by excessive concern about food and body shape and weight. Cardiovascular complications of AN may be life threatening. Reduction in the Coronary Blood Flow (CBF) can lead to ischemia, valve regurgitation, cardiomyopathy, and arrhythmias. The purpose of our study was to characterize the CBF in adolescents with AN.
Methods: Coronary flow velocities (peak diastolic (Vd) and systolic (Vs)) in the left anterior descending (LAD) coronary artery were prospectively studied with 2D/pulsed Doppler ultrasound in adolescents with AN during their malnourished state. Control group consisted of healthy age and sex matched adolescents. Significance was set at $p < 0.05$
Results: 42 adolescents with AN were included; 11/31 males/females, mean age: 15.3 ± 2.4y, mean weight loss: $22.3 \pm 11\%$, mean admission BMI 16.8 ± 2.9Kg/m2, median percentile BMI for age and gender $84.5 \pm 13.3\%$, minimal nocturnal heart rate of 38 ± 6 beats per minute (bpm). 28 healthy controls were compared to the AN group. LAD Peak Vd was 23.3 ± 7.5 cm/sec in the AN group versus 35.9 ± 8.9 cm/sec in the controls and peak systolic velocity was 11.7 ± 5 in AN and 18.4 ± 4.5 in the controls; $p < 0.00001$. Reduced CBF was found to be correlated to lower BMI percentile ($p = 0.009$), nocturnal bradycardia ($p = 0.039$) and diurnal bradycardia ($p = 0.003$). Anorectic males were significantly more medically unstable than the females (nocturnal bradycardia of 34.3 ± 4.7bpm versus 39.1 ± 5.7bpm, hospitalization days for reaching minimal heart rate of 45: 16.1 ± 5.2 Vs. 10 ± 5 days), yet no statistically significant difference was found in coronary flow between the genders.
Conclusions: A significant decrease in LAD diastolic and systolic flow velocities was found in adolescents with AN, and to the best of our knowledge is the first report of this potentially hazardous complication that may lead to papillary muscle dysfunction, valve regurgitation, arrhythmia and potential cardiomyopathy.

P2560 - LATE PNEUMOCCOCAL ENDOCARDITIS OF ATRIAL SEPTAL OCCLUDERS

Liat Gelernter- Yaniv[1], Mordechai Pollak[2], Isaac Srugo[3], Ellen Bamberger[4], Dawod Sharif[5]
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Late (>12months post implantation) bacterial endocarditis (SBE) following trans-catheter atrial septal defect (ASD) closure is rare. SBE prophylaxis is recommended for 6-12 months to allow full endothelialization. We present a patient with ASD occluder endocarditis and review the literature using PUBMED database. A 16-year-old obese male presented with 5-days of frontal headaches and fatigue. He had trans-catheter patent ductus arteriosus occlusion and ASD closure by Amplatzer device at 4-years of age. He was afebrile, tachycardic and tachypneic, had elevated CRP and a positive culture for intermediately-resistant Streptococcus-pneumoniae, subtype 17 f. A transesophageal echocardiogram showed a hypermobile 1X1.5 cm mass compatible with vegetation, at the left lower posterior edge of the ASD occluder (fig.1) and no residual shunt. The patient underwent extraction of the occluder and vegetectomy, followed by ASD closure. PCR from the vegetation was positive for S. pneumoniae. Notably, the occluder was completely endothelialized. The patient received 6 weeks of broad-spectrum antibiotics and fully recovered.
Results: 7 cases of late SBE of ASD occluders have been published (table 1). Only two in children. The organism was MRSA in 3, MSSA in two, unknown in two. Only our patient had pneumococcal endocarditis. In only two patients (including ours), the device was fully endothelialized.
Discussion: Late endocarditis of ASD occluders is exceedingly rare. Sporadic cases were mostly attributed to Staphylococcus species. This is the first report of Pneumococcal endocarditis. Incomplete

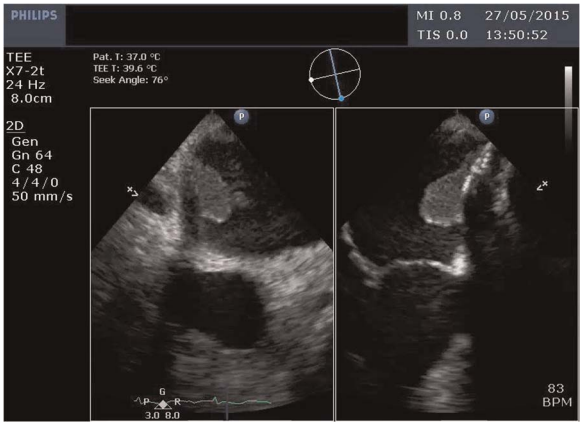

Figure 1.

Table 1. Cases review from the literature regarding late ASD occluders' endocarditis.

#	age	sex	defect	Device type**	Residual shunt	Time after implantation	Micro-organism	management	Endothelialization state	Risk factors	Presenting symptoms	published
1	10	f	ASD	ASO	?	6 years	unknown	Removal of device + abx	Incomplete endothelialization		Fever, heart failure, pleural effusion	World J Cardiol 2015
2	37	m	ASD - secundum	24mm ASO	no	4 years	MSSA	Removal of ASO + 6 w of nafcillin + rifampin *	Incomplete endothelialization on left atrial surface of mesh	Periodontal scaling w/o prophylaxis	Fever, myalgia, janeway lesions	Circ. 2015
3	37	m	PFO	ASO	?	2 years	MRSA	Removal of ASO + 6 w abx	Completely endothelialized	Obesity, DM type 1	Chest pain, diaphoresis, malaise	AnnThorac Surg 2014
4	59	m	ASD - secundum	ASO	?	12 years	MRSA	6 w of daptomycin (w/o removal of ASO)	?	HTN, DM type 2, CHF, s/p implantable cardioverter defibrillator	Sepsis	Indian Heart J. 2013
5	49	f	PFO	Helix occluder device	no	8 years	unknown	Removal of device + abx	?	?	Fever, malaise, possible embolic event	Echo cardiography 2011
6	66	m	ASD - secundum	32mm ASO	?	30 months	MRSA	Removal of ASO + 6 w of vancomycin	Poorly endothelialized	CAD, HTN, DM, hyperlipidemia, AF	fever	Am J Cardiol 2010
7	4	f	ASD - secundum	22mm ASO	?	12 months	MSSA	Removal of ASO + 6 w of nafcillin *	Minimal endothelialization	Mild mitral valve prolapse	Septic shock	Circ 2008

ASO = Amplatzer septal occlude
* Synergistic gentamicin treated initially; ** all devices were administered with trans catheter access.

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device endothelialization and residual shunt are well-established risk factors for endocarditis; this patient's device was completely endothelialized with no shunt.
Conclusion: although rare, endocarditis of ASD occluders must be considered. Regarding the number of new cases of device related endocarditis, long term follow-up and a high index of suspicion in patients with intra-cardiac devices is necessary. Guidelines for antibiotic prophylaxis should be reconsidered if more cases of late occluders endocarditis are reported.

P2565 - TYPE 1 TRUNCUS ARTERIOSUS WITH POSTERIOR SINUSAL ORIGIN OF THE MAIN PULMONARY ARTERY AND LEFT CORONARY ARTERY

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Princess Margaret Hospital for Children, Cardiology, Subiaco-Australia[1]; Lady Cilento Children's Hospital, Cardiac Surgery, South Brisbane-Australia[2]

Introduction: Truncus arteriosus (TA) is an uncommon congenital heart defect with a solitary ventriculo-arterial connection giving rise to the systemic, pulmonary and coronary circulations. We present a unique case of TA with the pulmonary artery originating from the posterior truncal valve sinus.
Case Report: A 23-year-old woman was referred for fetal echocardiography at 22 weeks gestation following an abnormal obstetric ultrasound. A common arterial trunk overriding a large ventricular septal defect (VSD) with proximal origin of the main pulmonary artery (MPA) was identified. A diagnosis of TA was made. Amniocentesis demonstrated 22q11 microdeletion. A female infant weighing 2850 g was delivered by normal vaginal delivery at 39 weeks gestation. She was mildly cyanosed with saturations of 85%. Transthoracic echocardiogram confirmed the diagnosis of TA. A solitary trileaflet truncal valve with trivial incompetence was overriding a large VSD. The MPA arose below the sinotubular junction from the posterior truncal sinus, with confluent left and right branch pulmonary arteries. During systole, the pulmonary orifice was partially occluded by the posterior cusp. The left coronary artery (LCA) arose anomalously from the posterior sinus, close to the anterior-posterior commissure. No left anterior descending artery was identified: the LCA coursed along the atrioventricular groove as the circumflex artery. The right coronary artery (RCA) arose from the centre of the anterior sinus. Cardiac CT angiogram confirmed the echocardiographic findings and demonstrated a left anterior descending coronary artery arising from the RCA. Successful surgical repair occurred on day 18 of life.
Conclusion: This anatomy is unique given the origin of the pulmonary trunk below the sinotubular junction from the posterior truncal sinus. There are no previous reports of posterior sinus origin of the common pulmonary orifice in Type 1 TA. Early identification of anatomical variants is essential in the successful surgical management of such infants.

P2570 - TEE DIAGNOSIS OF RIGHT ATRIAL THROMBUS IN A CASE OF CHRONIC CONSTRICTIVE PERICARDITIS

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Background: Chronic constrictive pericarditis is a debilitating condition with tuberculosis being the major cause in developing countries. Dyspnea and congestive symptoms are the common complaints at presentation requiring further evaluation and confirmation by imaging and clinical data. Association of right atrial thrombus with constrictive pericarditis is rare but needs to be ruled out as it can dislodge during manipulation or cannulation for cardiopulmonary bypass potentially causing massive pulmonary thromboembolism.
Material and Method: A 14 year old male presented with history of abdominal distension and pedal edema of 5 months duration. Physical examination revealed raised jugular venous pulse with sinus rhythm, normal heart sounds and no murmurs. Transthoracic echocardiography revealed constrictive physiology and thickened pericardium. Transesophageal echocardiography revealed an irregular hyperechoic mass of maximum dimensions 50x36 × 30mm in the right atrial free wall which was not detected on transthoracic examination and computed tomography. Pericardectomy followed by surgical removal of the thrombus using cardiopulmonary bypass was performed and the patient was discharged home after an uneventful course.
Result: Endocardial damage and intracardiac blood stasis secondary to decreased diastolic compliance can cause thrombus formation in constrictive pericarditis. Tranesophageal echocardiography in addition to confirming the pericardial pathology revealed the thrombus which was not detected earlier. Calcified pericardium distorting the right ventricle obscured the mass when transthoracic echocardiography was done from the apical window, while it not could be differentiated from the thickened pericardium on computed tomography. Right atrial thrombus can be associated with tricuspid valve obstruction, systemic and pulmonary thromboemboli and has an 4% early mortality rate warranting surgical removal under cardiopulmonary bypass. BIcaval cannulation was necessary in our case to avoid dislodging the thrombus.
Conclusion: Transesophageal echocardiography was instrumental in our case in the diagnosis of the thrombus resulting in a major change in surgical plan thus averting significant morbidity and possibly mortality.

P2573 - APICAL NON COMPACTION ASSOCIATED WITH CONGENITAL HEART DISEASES DIAGNOSED BY ECHOCARDIOGRAPHY

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Background: Isolated left ventricular non-compaction (LVNC) is reported extensively. But apical non-compaction (ANC) of both the ventricles and inter ventricular septum (IVS), is not reported much in literature as it is said to be common in reptiles but rare in human beings. Aim of our study is to analyze and evolve the echocardiographic (TTE) diagnostic criteria for "apical non-compaction ".
Material and Results: From January 2011 to February 2016, 100 consecutive cases that fulfilled standard echocardiographic criteria for LVNC and the additional criteria 1) Swiss cheese appearance of IVS, looking like a delta of the river 2) non-compaction of right ventricle on TTE, formed material for this study. Age ranged from 3 days to 35 years, with 37 males and 33 females. The associated lesions were present in all almost all cases. 86 patients had acyanotic heart disease (86%) and 14 patients had cyanotic heart disease (14%). 21 of 100 cases had pump failure (21%). 10 cases had LV

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dysfunction, 8 had RV dysfunction and 3 had biventricular dysfunction, 38 patients (38%) had pulmonary hypertension; two had thrombus. 3 cases of Swiss cheese VSDs were post-operative residual shunts. ANC is known to happen in the chicken heart, but thus far there has been no evidence to suggest a similar mechanism in human beings. ANC was totally ignored by the echocardiographer or only specific lesions like Swiss cheese VSD were diagnosed in the past. For the first time we report a large series of 100 cases of ANC diagnosed by TTE only
Conclusion: TTE with color Doppler is an excellent modality to diagnose ANC with additional criteria. Probably ANC is ignored. ANC is invariably associated with other serious congenital cardiac malformations, which worsen the pump failure.

P2578 - ROLE OF CONTRAST ECHOCARDIOGRAPHY WITH AGITATED SALINE INJECTION FOR ANTEMORTEM DIAGNOSIS OF CORTRIATRIATUM DEXTER

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Background: Cor triatriatum dexter (CTD) is an extremely rare congenital anomaly (0.025%), in which the right atrium (RA) is divided into two chambers by a septum. Anomaly is usually diagnosed either on autopsy or during surgery as diagnosis is missed clinically. We report a series of 16 cases of CTD diagnosed antemortem by 2D transthoracic echocardiography (TTE) with contrast echo.
Objective: To describe varied clinical presentations and highlight importance of TTE with contrast prepared with patient's blood agitated with 0.9% saline, injected from upper and lower limbs for precise antemortem diagnosis of CTD.
Material, Methods and Results: Sixteen consecutive patients of CTD diagnosed with TTE, formed the material. Age ranged from 2 days to 51 years. All patients underwent detailed TTE interrogation especially in RV inflow view with agitated saline contrast injected from both upper and lower limbs separately to demonstrate membrane in right atrium. Two cases referred as primary pulmonary hypertension (PPH), another two with hemoptysis and pulmonary embolism turned out to be CTD. One had multiple thrombi in right atrial appendage. Seven patients had atrial septal defect (ASD). 2 days old neonate with severe cyanosis(SPO2-54%) died, as both SVC and IVC were opening in proximal chamber and desaturated blood was shunting to left atrium. 9 days old Ebstein's anomaly with ASD, the CTD was suspected, prenatally by fetal echo. CTD is not always be benign, as described in the literature, because it was fatal in 3 cases (20%) in our series.
Conclusion: Ante mortem diagnosis of CTD with agitated saline contrast echocardiography can be precise and lifesaving. CTD should be looked for in RV inflow view in all cases of Ebstein's anomaly and ASD, who present with PAH or pulmonary embolism.

P2580 - RIGHT VENTRICULAR FUNCTION ASSESSED BY THREE DIMENSIONAL ECHOCARDIOGRAPHY AFTER CONE PROCEDURE COMPARISON WITH MAGNETIC RESONANCE IMAGING

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Background: Assessing right ventricular (RV) systolic function using echocardiography is challenging due to its complex morphology. Although cardiac magnetic resonance (CMR) is the "gold standard" to evaluate RV, three dimensional echocardiography (3D Echo) has been shown to be an alternative to accurately evaluate cardiac function. We aimed to compare parameters of RV function by two dimensional echocardiography (2DEcho) and 3DEcho to CMR in patients with Ebstein´s anomaly after surgical treatment with Cone procedure.
Methods: We assessed patients during late post-operative follow-up (median 7 years, range 1-21 years) from Cone procedure regarding clinical status, 2DEcho RV parameters including TAPSE, fractional area change (FAC), tissue Doppler S velocity, Tei index and longitudinal Strain and RV volumes and ejection fraction (EF) using 3DEcho and CMR (performed within 4 hours of echocardiography). Pearson´s correlation was used for analysis.
Results: Out of 27 patients (13 male, median age 20 years, range 8-54 years), 2 were excluded from analysis due to inadequate echocardiographic window. All patients were in regular cardiac rhythm. Most patients (44,4%) complained of effort dyspnea. None of 2DEcho parameters showed a good correlation to EF using CMR. Pearson's correlation between 3DEcho and CMR was good for RV volumes and moderate for EF (Table 1).
Conclusion: Compared to CMR, 3DEcho was able to adequately evaluate RV systolic volumes and function in operated Ebstein's anomaly. Systolic function parameters from 2DEcho did not correlate well to CMR. Despite good surgical results, RV function remained impaired in this specific set of patients.

Table 1. Comparison of RV volumes and EF between 3DEcho and CMR.

	3DEcho	CMR	r	p
RV end diastolic volume	126 ml (60-325)	177 ml (76-423)	0.7	<0.001
RV end systolic volume	87 ml (265-33)	101 ml (41-334)	0.8	<0.001
EF	32% (19-54)	41% (21-53)	0.6	<0.001

P2581 - 3D ECHO EVALUATION OF RIGHT VENTRICULAR VOLUMES IN THE LATE POSTOPERATIVE FOLLOW UP OF TETRALOGY OF FALLOT REPAIR WITH PULMONARY INSUFFICIENCY

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Introduction: Right ventricular (RV) functional and volumes evaluation is one of the major concerns in the follow-up of patients who underwent tetralogy of Fallot (ToF) repair. Chronic pulmonary insufficiency (PI) results in RV dilation and dysfunction. Timing for re-intervention has been defined mainly by RV diastolic and systolic indexed volumes (RVEDVi and RVESVi) assessed by magnetic resonance (MRI). The specific RV 3-D echo

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Tomtec@ software is an advanced tool that allows calculation of the same parameters and may be applied in this specific population. This study aimed to test the correlation between RVEDVi and RVESVi of patients with PI in the late follow-up of ToF repair assessed by MRI and 3-D echo.
Methods: 30 patients were enrolled in the study. Patients with moderate to severe residual lesions except for PI were excluded. 3-D echo was performed in our echo lab and the MRI in outside hospitals.
Results: Time between surgery and study was 18 years (6–44 years). Time between the 2 studies was 1 year (1 month – 4 years). PI was moderate in 13% and severe in 70% of the cohort. The correlation of the RVEDVi and RVESVi between the two images methods was $r = 0{,}7 - p < 0{,}001$ and $r = 0{,}68 - p < 0{,}001$ respectively with greater discrepancies observed in larger volumes. RVEDVi $\geq$ 99 ml/m^2 had sensitive and specificity values of 69 and 93% respectively in estimating RVEDVi $\geq$ 140 ml/m^2 measured by MRI.
Conclusion: There is reasonable correlation between RMI and 3D-echo in the assessment of RV volumes. It seems that 3D-echo may be an option for the regular monitoring of the RV volumes in the late postoperative period of ToF repair until critical volumes are reached, when MRI should be performed to identify the optimal timing to intervene in the RV outflow tract.

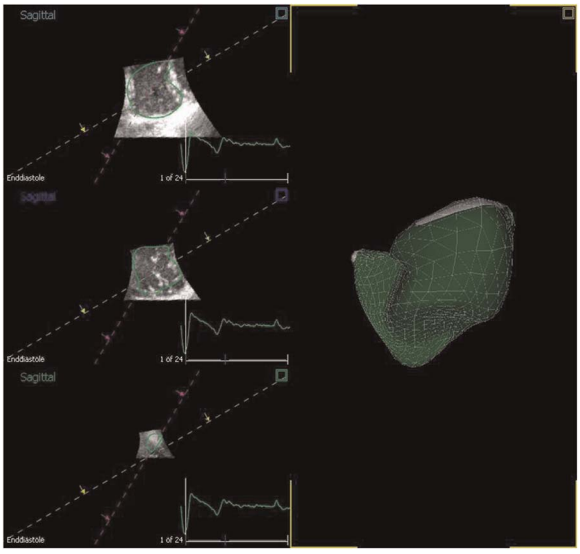

Figure.

P2594 - UNEARTHING THE UNIQUENESS OF CONGENITAL HEART DISEASE CREATIVE PARTICIPATORY PRACTICES CARDIOVASCULAR MAGNETIC RESONANCE IMAGING AND 3D PRINTING

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Background: Patients with congenital heart disease (CHD) require life-long follow-up. Adolescents transitioning from the pediatric to adult centre need to gain an appreciation of their cardiovascular anatomy and the lifestyle implications of their CHD. We propose that an interdisciplinary approach merging medical imaging, 3D printing technology and artistic methodologies could be beneficial for CHD patients to get in touch and engage with the uniqueness of their condition.
Methods: CHD patients (n = 14, 14-18 years, 6 males) participated in one of two, two-part, creative workshops (7 young people in each) facilitated by an artist, a health psychologist, a bioengineer and an adolescent nurse specialist. The two sessions were run 1.5 months apart. Creative exercises included blindfolded drawing and sculpting self-portraits, creative writing, body mapping and metal embossing. These activities facilitated the development of imagery and language to explore individual uniqueness. Patient-specific heart images were derived from the participants' MRI data. MRI-derived patient-specific heart models were manufactured by means of 3D printing. Heart images were re-interpreted artistically and 3D models were discussed, in an effort to explore heart-related imagery and narratives.
Results: CHD patients engaged extremely well with the inter-disciplinary process. The creative output was charged with experiential and medical connotations, and was articulate and confident. Patients found 3D vs. 2D a more immediate form of expression. Heart narratives were potent and unique, including eloquent metaphors of survival, resilience, strength, fragility and structural complexity. 3D heart models aided the conversation around anatomy and understanding it. Participants' feedback highlighted enjoyment, the opportunity to get in touch with one's self, the possibility of working with peers and the privilege of reflecting on uniqueness.
Conclusions: An interdisciplinary approach, including artist-led workshops and MRI-derived models, can aid adolescent CHD patients to explore individual uniqueness as well as the form and significance of their own heart.

P2600 - QUANTITATIVE EVALUATION USING LUNG PERFUSION SCINTIGRAPHY FOR FONTAN CIRCULATION

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Background: After the Fontan procedure, a right-to-left (R-L) shunt by venous-venous shunt or pulmonary arteriovenous fistula could develop, which causes hypoxemia. In the present study, we tried to evaluate the R-L shunt in patinets with fontan procedure on lung perfusion scintigraphy.
Subjects: Total 25 patients (13males, 12females; age,2.4-25.3years) who underwent lung perfusion scintigraphy after Fontan procedure, respectively.
Methods: 60% amount of isotope 99mTc-MAA was injected into lower extremities and the other 40% amount of isotope was injected into upper extremities. The R-L shunt rate ((whole body count - lung count)/whole body count x 100%) was calculated and was compared with hemodynamic date from simultaneous cardiac catheterization.
Results: the R-L shunt rate and arterial oxygen saturation (SpO2) showed a linear, reverse correlation ($Y = -0.26x + 98.2$ and $r = 0.78$, $p < 0.01$). Furthermore, in comparison between high central venous pressure (CVP) group (≥12mmH) and low CVP group (<12 mmHg), the R-L shunt in high CVP group was significantly higher than that in low CVP group ($36.2 \pm 17.5\%$ vs. $17.8 \pm 9.9\%$, respectively, $p < 0.01$). There was not statistically significant difference between the R-L shunt rate and pulmonary vascular resistance or postoperative period.

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Conclusion: Quantitatively evaluatation of the right to left shunt using lung perfusion scintigraphy is feasible and useful after Fontan procedure.

P2608 - CONGENITAL DOUBLE LUMEN AORTIC ARCH A RARE ANAMOLY

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Background/Hypothesis: Persistence of fifth aortic arch is extremely rare anomaly presenting as double lumen aortic arch.
Material and Methods: Case report.
Results: We report a case of double lumen aortic arch best explained by persistence of left fifth aortic arch. It was associated with membranous ventricular septal defect. The patient underwent successful repair of septal defect, while arch anomaly was asymptomatic and left untreated.
Conclusion: Double lumen aortic arch is extremely rare anomaly due to persistence of left fifth aortic arch, if asymptomatic, does not require any treatment.

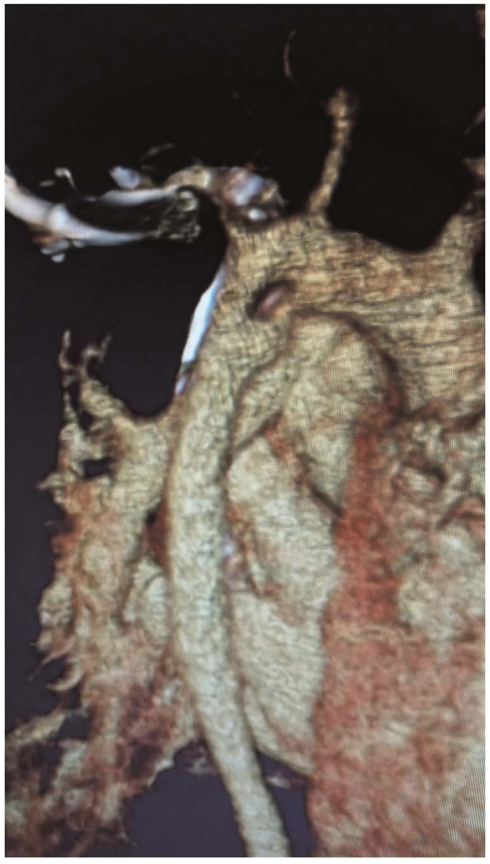

Figure.

P2619 - POSTNATAL FINDINGS IN RIGHT TO LEFT HEART SIZE DISPROPORTION IN FETAL ECHOCARDIOGRAPHY – A GENERAL HOSPITAL EXPERIENCE

Claudia Gallafrio[1], Samira Morhy[1], Alessandro Lianza[1], Glaucia Tavares[1], Rita Sanchez[2], Wercules Oliviera[1], Tania Afonso[1], Ana Rodrigues[1], Marcelo Vieira[1], Claudio Fischer[1]
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Background/Hypothesis: In fetal echocardiography, right to left heart size disproportion, with the right heart larger than the left (R-L/HD), may be associated with left-side obstructive disease, such as coarctation of the aorta (CoA). These findings isolated are nonspecific for predicting postnatal diagnose of CoA, and often need repeated echocardiograms, leading to significant parents stress. The objective of this study is to identify associations R-L/HD and left-sided obstructive disease.
Methods: From April 2011 to February 2016, 64 out of 1555 fetal echocardiograms were identified with isolated L-R/HD, with 18 delivered in our hospital. Pre and postnatal echocardiograms and the medical records were reviewed. Medium gestational age was 31 weeks (25 to 36). Echocardiography measurements included: tricuspid/mitral valve diameters, left/right ventricle mid-cavitary width dimensions, pulmonary/aortic valve diameters, pulmonary/aorta dimensions, transverse aortic arch and aortic isthmus diameters. Flow directions at atrial level and across aortic and ductal arches were analyzed.
Results: Two fetuses presented with arch obstruction, one of them critical, requiring neonatal intervention. Another presented with isolated quadricuspid aortic valve. None of the parameters distinguished critical CoA from a normal heart, though there was a tendency for the aortic isthmus diameter to be smaller in fetus with CoA(2,3mm vs 3,8mm respectively),
Conclusion: In our institution, most cases with isolated R-L/HD in fetal echocardiography presented with normal hearts after birth, suggesting this might not be an accurate means of antenatal detection of CoA.

P2621 - ASSESSMENT OF LINEAR INSERTION OF ATRIOVENTRICULAR VALVES IN FETUSES BY POSTMORTEM CARDIAC MRI

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Background: Linear insertion of the atrioventricular valves (LIAVV) on foetal echocardiography was described in 2002 by Fredouille in association with aneuploidy, and could represent a minor form of atrioventricular septal defect (AVSD). However, no anatomic or embryologic explanation has been provided to confirm this hypothesis. To elucidate the pathological significance of LIAVV, we compared postmortem high-spatial resolution cardiac magnetic resonance imaging (MRI) with foetopathological examination for visualizing LIAVV in human fetal hearts. This work will be a preliminary step for a larger scale study, with as a final aim to confirm or not whether LIAVV belongs to the spectrum of AVSD.
Material and Methods: Two normal hearts (N), 1 AVSD and 2 LIAVV (gestational age 17-34 weeks) were randomly selected among the anatomic collection of the French Reference Center for Complex CHD. Postmortem cardiac MRI was performed with a 4.7-T imager. Specimens were rinsed and placed in airtight bags filled with 1-2 ml of distilled water. 3D and 2D Turbo-RARE sequences in four-chamber, short axis and left ventricular long-axis planes were performed with a minimal isotropic or

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in-plane resolution of 156 μm. MR images were compared to foetopathological examination. Quantitative analysis included measurement of the distance between the insertion of the medial leaflets of the tricuspid and mitral valves, and of the left ventricular inlet/outlet distance ratio.
Results: MRI and pathology findings agreed perfectly in identifying valve insertion, evaluating mean mitral valve-tricuspid valve distance (N: 1.63 mm; LIAVV: 0.89 mm) and inlet-outlet distance ratio (N: >0.9, LIAVV: < 0.80, AVSD: <0.5). Both LIAVV had a deficiency of the inlet ventricular septum closed by the insertions of a bi-leaflet mitral valve, explaining the lack of offsetting of the AV valves.
Conclusions: Postmortem cardiac MRI provides results similar to pathology for assessing LIAVV in fetuses without requiring specific preparation of the heart.

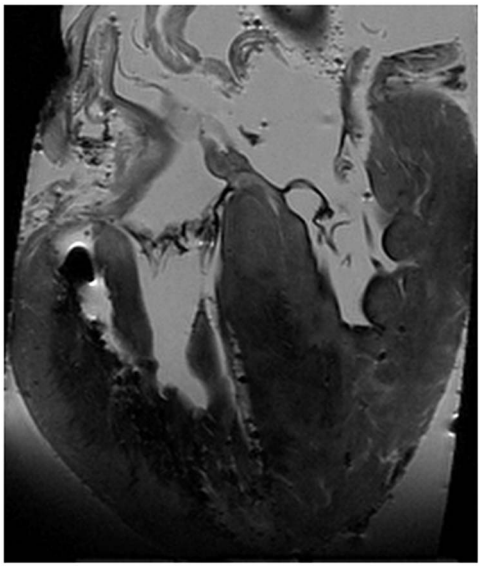

Figure.

P2628 - EBSTEIN'S ANOMALY AND MORPHOLOGY VARIATION (A ROLE OF PEDIATRIC CARDIOLOGIST IN FETAL ECHOCARDIOGRAPHY)

Syarif Rohmi[1], *Winda Azwani*[1], *Gatot Abdurrazak*[2], *Poppy Roebiono*[3], *Dicky Fachri*[4]
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Background: Ebstein's Anomaly (AE) is a rare congenital cardiac defect and characterized by remarkable morphologic variability, a broad spectrum of clinical presentations and treatment. Detailed fetal echocardiography plays important rule to make an accurate diagnosis. During 2012-2016, there were 4 AE cases, 1st three cases easily to diagnosed. The 4th case was referred to evaluate the diagnosis. Here, we present the 1st and the last case.
Case 1: 25 years healthy mother, G2P1AoH1, full gestational age, estimated weight 3200 gr, referred for reevaluation Ebstein's Anomaly. Four chamber view (4CV): very dilated RA and RV, severe TR, severe downward septal leaflet tricuspid valve (TV), PFO and smallish LV. Five chamber and trachea view: normal. Normal PA, PFO, small PDA and left aortic arch.
Case 4: 24 years full gestational healthy mother with very dilated RA and RV, severe TR, slight smallish RV compare to normal LV dimension on 4 CV. Further detailed examination revealed severe downward TV septal leaflet, severe TR and PFO. Five chamber and trachea view were transposed Great arteries, PFO and PDA. Diagnosis was severe Ebstein's anomaly, transposed GA (?), PFO and PDA.
Management: Case 1. Nasal CPAP, flow 5, FiO2 0.6. Laboratories: normal, SaO2 76-88%. Patient underwent Starness procedure and patient died on 6th day due to heart failure. AS 9/10 Case 2. Underwent caesarean delivery and transferred to NICU, spontaneous breathing. TTE: EA, ccTGA, large PDA and postductal coarctation of aorta (?).
Laboratories: normal. Team decided to start PGE1 with SaO2 68%. MSCT was planed and ongoing discussion to decide the appropriate procedure.
Conclusion: Detailed Ebstein's anomaly fetal echocardiogram is very important to assess accurate diagnosis for early treatment and intervention.

P2632 - A UNIQUE CASE SERIES OF CHILDREN WITH CONGENITAL PULMONARY VEIN STENOSIS FROM AN AFRICAN CENTER

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Background: Congenital pulmonary vein stenosis (PVS) is a rare condition, which results from abnormal embryological incorporation of the common pulmonary vein into the left atrium.
Methods: A retrospective descriptive case series study was conducted with the aim of describing the characteristics and outcome of children with congenital pulmonary vein stenosis at an African tertiary care center over a 25-year period. A Computerized paediatric cardiology database initiated in the early 1990s was sourced to identify patients whose hard copy clinical notes were retrieved and reviewed.
Results: Five cases of congenital PVS were identified between January 1990 and January 2016. Congenital PVS accounted for 0.07% of all congenital heart defects seen at the center during the study period. The age at diagnosis ranged from 22 months to 13 years. The majority of patients manifested with respiratory symptoms, with two patients presenting with recurrent haemoptysis. The diagnosis of PVS was confirmed by cardiac catheterization and pulmonary angiography in all patients. All cases were right-sided unilateral PVS and all were associated with one or more congenital heart defects. Mild pulmonary hypertension and elevated capillary wedge pressures were found in all patients. Only one patient underwent specific surgery to relieve the PVS, which subsequently recurred. The other patient operated on was for lobectomy following recurrent haemoptysis, this patient subsequently died of sepsis.
Conclusion: Congenital pulmonary vein stenosis is a rare condition often associated with other congenital cardiac defects. Respiratory symptoms are common at presentation, with haemoptysis forming part of the presenting clinical spectrum, in keeping with the published literature.Moreover, elevated pulmonary artery pressures and increased ipsilateral pulmonary capillary wedge pressures are a common finding at catheterization.

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P2640 - MELODY VALVE PLACEMENT FOR MITRAL VALVE DYSPLASIA SYNDROME

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Background: Mitral valve dysplasia syndrome includes a dilated left ventricle and a dysplastic mitral valve often necessitating repair or replacement. Options for neonatal mitral valve palliation are challenging, but the placement of Melody mitral valve shows promise in this patient population.
Case: A 6 week old female, born full term via an uncomplicated pregnancy, had normal prenatal ultrasounds. After birth, she failed the pulse oximetry test prompting a cardiac evaluation. The echocardiogram showed severe mitral valve regurgitation and a dilated left ventricle and atrium. She was medically managed and initially sent home but returned to the referring hospital with difficulty feeding and respiratory distress requiring intubation.
Decision-making: On review of her records, her echocardiogram was consistent with mitral valve dysplasia syndrome with severe mitral valve regurgitation. There was an arcade type defect of the mitral valve which was unlikely to improve with medical therapy alone. Alternative options included a heart transplant, primary repair and a prosthetic mitral valve, which would require anticoagulation. However, given her size and the lack of small mitral valve prostheses, she was not a candidate for the aforementioned options. After extensive case discussion, decision was made for placement of a stent mounted by prosthetic Melody valve in the mitral position, performed surgically. She tolerated the procedure well and her echocardiogram prior to discharge showed a well-functioning Melody valve in the mitral position with no stenosis or regurgitation. To date, there have been no episodes of respiratory distress, poor perfusion or syncope. Follow up echocardiograms have showed no significant mitral valve regurgitation or left ventricular dysfunction.
Conclusion: This case highlights the feasibility of a Melody valve placement in the mitral position, as a new option for infants with a dysplastic mitral valve.

P2641 - ACUTE MYOCARDITIS WITH INFARCT LIKE PRESENTATION IN A PAEDIATRIC POPULATION. ROLE OF CARDIOVASCULAR MAGNETIC RESONANCE

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Vega Baja Hospital, Pediatric Cardiology, Orihuela-Spain[1]; Vall D'hebron Hospital, Pediatric Cardiology, Barcelona-Spain[2]; Vall D'hebron Hospital, Pediatric Radiology, Barcelona-Spain[3]; Josep Trueta Hospital, Pediatric Cardiology, Girona-Spain[4]; Sant Joan De Reus Hospital, Pediatric Cardiology, Tarragona-Spain[5]

Background: Chest pain is a typical symptom of acute myocarditis in teenagers and young adults. It may be indistinguishable from myocardial ischemia so it is called "infarct-like pattern". Cardiovascular magnetic resonance has an important role as a non-invasive diagnostic tool. The aim of our study is the description of an acute myocarditis series with infarct-like pattern, and the evaluation of the cardiovascular magnetic resonance role in a paediatric population.
Material and Methods: We included all paediatric patients (0-16 years) admitted to our hospital (May 2007 - May 2016) with clinical diagnosis of acute myocarditis and infarct-like presentation (chest pain, ECG alterations and elevated cardiac biomarkers). Data about patient demographics, test results and outcomes were analyzed.
Results: We collected 7 patients (5 males, 2 females) with a median age of 14 years (12.5-15.2) were included. All patients showed ST-segment changes and elevated cardiac biomarkers. Three patients had left ventricular hypertrophy (50%) and 2 presented mild systolic left ventricular dysfunction (33.3%). All patients had at least 2 positive Lake-Louise criteria. Late gadolinium enhancement was positive in all of them. With a median follow-up of 23 months (8-47), all of them are alive, with no cardiac symptoms and free of heart transplant.
Conclusions: Chest pain is a typical symptom in teenagers with acute myocarditis. If it is suspected, cardiovascular magnetic resonance should be performed as a first line diagnostic tool. Unlike what was described in adults, late gadolinium enhancement does not imply worse outcome in teenagers.

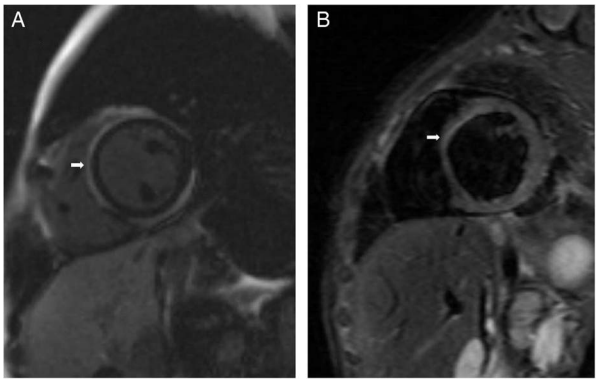

Figure.

P2653 - CLINICAL RADIOLOGICAL AND PATHOLOGICAL CORRELATIONS REVEAL A PATTERN OF INCREMENTAL BRAIN INJURY AND IMPAIRED CEREBRAL GROWTH IN PATIENTS WITH CONGENITAL HEART DISEASE

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Background: Impaired brain growth and white matter injury (WMI) are frequently seen on MRI in newborns with congenital heart disease (CHD) both before and after surgical treatment. However there is limited information available regarding the relationships between clinical, imaging and pathological findings in this setting. Here we report on three cases in which autopsy with the examination of the brain was performed from a larger cohort of children being prospectively studied with serial brain MRI.
Materials and Methods: Between 2013 and 2016, 95 newborns with CHD underwent brain MRI imaging both before and after surgery in our department. 44% had single ventricle physiology and 53% had transposition of great arteries. At a mean follow-up interval of 19 months, 11 patients had died (12%). Autopsy that included examination of the brain was performed in 3 patients (2.7%).

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Results: Subject 1 had double outlet right ventricle with mitral atresia, subaortic and aortic stenosis. Subject 2 had right isomerism with an atrioventricular septal defect and double outlet right ventricle with pulmonary stenosis and obstructed infracardiac total anomalous pulmonary venous return. Subject 3 had Tetralogy of Fallot with hypoplastic pulmonary arteries and Adams-Oliver Syndrome with a NOTCH 1 mutation. Mean pre-operative, post-operative, and post-mortem brain volume z-scores were -1, -2.5 and -2.9 respectively. In addition, there was evidence of punctate WMI following surgery but also present pre-operatively in Subject 3, and further watershed and embolic infarction following post-operative complications with hemodynamic instability. The key imaging and pathology findings are shown in Table 1.
Conclusions: CHD is associated with poor brain growth, which is present at birth but progresses following palliative surgery. While uncomplicated surgery is often associated with no new brain injury, periods of hemodynamic instability and cardiac arrest are frequently accompanied by further ischemic injury. As yet undefined predisposing genetic factors may influence the extent of the brain injury.

Table.

Subject Number & diagnosis	Age at preop brain MRI (days)	Preoperative MRI findings	Brain weight (z score)	Type of Palliative Surgery between MRI examination	Age at postop brain MRI (months)	Postoperative MRI findings	Brain weight (z score)
1.DORV with MA	3 d	Corpus Callosum: thin Ventricles: normal CSF spaces: normal Hemorrhage: none WMI: none Stroke: none	-0.4	I) PAB 2) Norwood-BT; 3) ECMO; 4) coarctation stent	2 m	Corpus Callosum: thin Ventricles: normal CSF spaces: normal Hemorrhage: few scattered foci of susceptibility WMI: none Stroke: none	-1.7
2. RAI, AVSD, DORV, PS, TAPVC	5 d	Corpus Callosum: thin Ventricles: normal CSF spaces: enlarged Hemorrhage: none WMI: none Stroke: none	-1.2	I) pulmonary vein stent 2) RVOT stent 3) Sutureless pulmonary vein repair	3 m	Corpus Callosum: thin Ventricles: normal CSF spaces: 7mm subdural collection, possibly subdural hygroma. Hemorrhages: multiple scattered foci of susceptibility; thin bilateral subdural hemorrhage. WMI: two foci of diffusion restriction in the genu of the right internal capsule and on the left centrum semiovale. Stroke: none	-2.7
3. TOF with severely hypoplastic PA, Adams Oliver Syndrome	44 d	Corpus Callosum: thin Ventricles: enlarged CSF spaces: normal Hemorrhage: none WMI: periventricular destructive changes, with bilateral calcifications. Stroke: none	-2.2	1) central shunt 3.5 mm and bilateral pulmonary plasty 2) central shunt 5 mm 3) removal of seroma and hematoma; 4) 16 mm RVOT conduit. VSD left open	15 m	Corpus callosum: thin Ventricles: increased ventriculomegaly CSF spaces: normal Hemorrhage: few scattered foci of susceptibility WMI: persistent destructive changes in periventricular and deep WM Stroke: none	-3

Subject Number & diagnosis	Clinical course and cause of death	Last brain imaging	Age of death	Pathological findings	Brain weight (z score)
1.DORV with MA	Postoperative course: Multiple LCOS Seizures, Cardiac Arrest	BrainMRI: Corpus callosum: thin Ventricles: enlarged CSF spaces: enlarged Hemorrhages: multiple scattered foci of susceptibility WMI: thin subcortical WM. Multiple bilateral areas of PVL within the watershed distribution of both cerebral hemispheres. Stroke: none Brain Volume: z score: -3.6	6 m	Corpus callosum: thin Ventricles: enlarged Hemorrhages: none WMI: thin subcortical WM .Multiple bilateral areas of PVL within the watershed distribution of both cerebral hemispheres. with areas of axonal swelling, cavitation and macrophage infiltration	-4
2. RAI, AVSD, DORV, PS, TAPVC	4) Redo sutureless repair, TV repair, main PA division with left BT shunt 4 mm Postoperative course: reduced ventricular function, hypotension, CPR, ECMO. Accidental cannula displacement: Cardiac Arrest. CPR. Dilated non responsive pupils	Brain CT angio: Abrupt occlusion of the distal basilar artery with an intraluminal thrombus and acute infarction of the left thalami Cardiac CT: Thrombus at the proximal part of the BT shunt at the anastomosis with the innominate artery	7 m (2 weeks after the stroke)	Corpus callosum: thin Ventricles: enlarged Hemorrhages: small hemorrhagic lesion in the left thalami. WMI: thin subcortical WM. Delayed myelination pattern. Stroke: subacute left thalamic infarction; basilar and right PCA thrombosis with recent large infarction in the right occipital lobe	-1.9
3. TOF with severely hypoplastic PA, Adams Oliver Syndrome	Sudden death at home. Thoracic hemorrhage at autopsy	NA	15 m	Corpus callosum: thin Ventricles: enlarged Hemorrhages: none WMI: thin subcortical WM. Multifocal WM changes consisting of myelin and axonal loss, microcalcification and macrophage infiltration. Remaining axons show swelling. Stroke: none	-3

AVSD:artiventricular septal defect; CSF: cerebrospinal fluid; DORV: double outlet right ventricle ECMO: extra corporeal membrance oxygenation; LCOS: low cardiac output syndrome; PAB: pulmonary artery banding; BT: Blalock Taussing; MA: mitral atresia; PCU: posterior cerebral artery PVL: periventricular leukomalacia; RAI: right atrial isomerism; RVOT: right ventricular outflow tract; TAPVC: total anomalous pulmonary vein connection; TOF: tetralogy of Fallot; WM: white matter; WMI: white matter injury.

P2656 - USEFULNESS OF MYOCARDIAL STRAIN IMAGING IN SPINAL MUSCULAR ATROPHY

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Background: Our aim was to evaluate left ventricular (LV) mechanics by using speckle tracking echocardiography (STE) in patients with Spinal Muscular Atrophy (SMA) in the presence of LV ejection fraction (EF) >50% and to compare it after 1 year of oral salbutamol.
Methods: We studied 51 patients with SMA (mean age 7.2 years; range 0-12 years, LVEF > 55%) and 59 age-matched normal controls (mean age 7.1 years; range 0-12 years). All underwent standard echocardiographic evaluation and STE.
Results: LVEF ($63.6 \pm 8.2\%$ vs. $64.1 \pm 5.1\%$, $p = 0.826$) was not different between patients and controls. Patients with SMA had higher heart rates and increased LV end-systolic dimensions compared with controls. Diastolic parameters were significantly abnormal in SMA patients versus controls (E/e' average: 11.9 ± 5.8 vs. 6.6 ± 3.0, $p = 0.0014$). Global LV longitudinal strain was significantly lower in SMA patients versus controls ($-17.6 \pm 3.5\%$ vs. $-23.4 \pm 3.1\%$, $p < 0.0001$). LV torsion ($9.1 \pm 4.9°$ vs. $11.9 \pm 3.3°$, $p = 0.046$) was significantly impaired in SMA patients. Diastolic parameters and global LV longitudinal strain values improved after 1-year-course of salbutamol in SMA patients ($p = 0.003$)
Conclusions: Diastolic function, LV longitudinal deformation and LV torsion are impaired in SMA patients compared to control group despite an LVEF >50%. A course of salbutamol improves all the previous parameters. We suggest including a detailed study of the diastolic function and cardiac mechanics in the clinical follow-up of these patients.

P2667 - COARCTATION OF AORTA AND VEIN OF GALEN ANEURYSMAL MALFORMATION IN A NEONATE

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Introduction: Galen ven aneurysm is a rare congenital intracranial vascular malformation. Aortic coarctation can be seen in Galen malformation. The reason is that the blood of the aorta is stolen by the brain due to the extensive malformation. We present a case of a newborn with coarctation of the aorta and a vein of Galen malformation (GM).
Case Report: A male infant was born spontaneously at 39 weeks. Echocardiography was performed on a patient who had a murmur on physical examination revealed a aortic coarctation, a PDA, and a PFO. Prostaglandin infusion was started and referred to our clinic for further treatment. Cardiac examination revealed 3/6 systolic murmur at the left sternal border. Pulses were described as strong in the upper and lower extremities. Repeated echocardiographic examination revealed severe aortic coarctation (figure 1) and large ductus with right to left shunt. There was also evidence of retrograde diastolic flow into the transverse aortic arch toward to head and neck vessels (figure 2). A brain computed tomograpy was also consistent with a very large vein of GM (figure 3). Interventional Radiology was consulted for vessel embolization; but the patient was referred to an advanced center because there was not enough equipment.

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Discussion: Twenty-three cases with congenital cardiac anomalies associated with GM were reported in the literature. Today's survival rate with endovascular treatment is 70-80% and full recovery is around 50%. In the case of coexistence of GM and aortic coarctation, descending aorta blood flow is completely ductal dependent. At the same time, the ductus also decomprese the pulmonary vascular bed. For this reason, GM should be repaired firstly in patients and then intervened in aortic coarctation. Otherwise, inevitable and intractable congestive heart failure will occur.

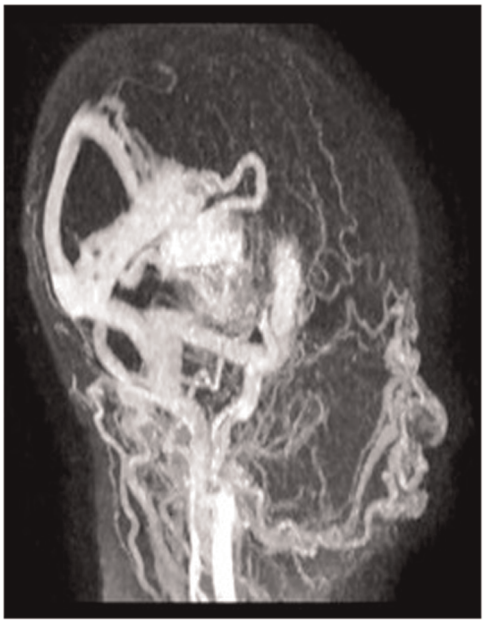

Figure 1.

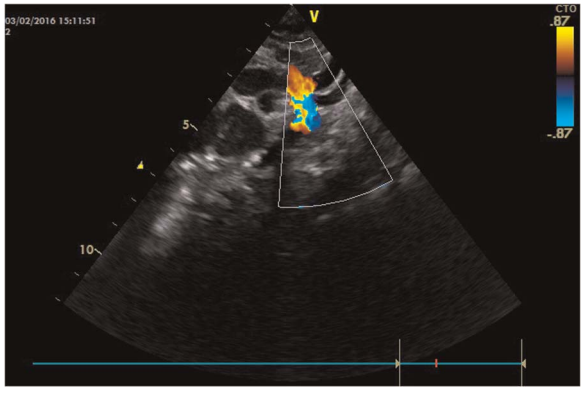

Figure 2.

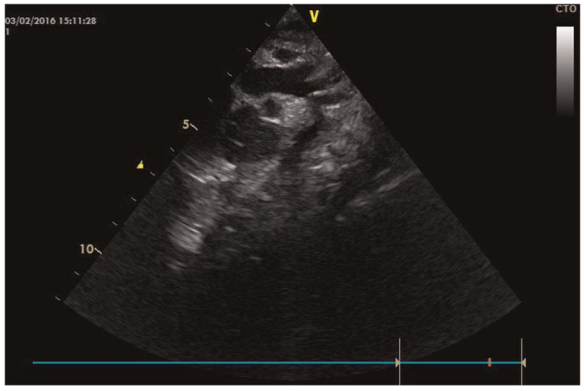

Figure 3.

P2699 - IMAGE COMPARISON OF CT AND ANGIOGRAPHY IN A 3 MONTHS OLD INFANT WITH ALCAPA AND HEART FAILURE

Chung-Chih Kao[1], Betau Hwang[2], Chun-Hsiang Ko[3], I-Hsin Lu[3], Shang-Yen Yang[4], Hua Guo[5]
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Background: ALCAPA(Anomalous Left Coronary Artery from the Pulmonary Artery) is a rare congenital coronary artery disease with high mortality rate,especially when congestive heart failure occurs in young infancy and should be differentiated from dilated cardiomyopathy.Better left coronary artery image would be helpful for differential diagnosis and treatment.
Materials and Methods: A 3 months old female presented with chief complaint of dyspnea for more than 3 weeks.Her chest X-ray showed cardiomegaly and EKG showed left ventricular hypertrophy and ischemic change with T-wave inversion in V4, V5 and V6.Echocardiogram revealed heart failure and a dilated right coronary artery but the left coronary artery was obscure.The patient underwent CT(computed tomography) scan under the impression of ALCAPA.Due to heart failure associated with sinus tachycardia,the volume of contrast became an issue.Diagnostic angiography was performed after CT scan of which images were inconclusive.
Results: CT scan showed fair right coronary artery images,but the origin of the left coronary artery is obscure and equivocal for anatomically the pulmonary trunk is very much close to the ascending aorta in young infancy.The ascending aorta angiogram showed a single coronary artery from the right coronary cusp. Selective right coronary artery angiogram showed collaterals to the left coronary artery and outflow to the pulmonary artery confirming the diagnosis of ALCAPA.
Conclusions: CT scan is fast for acquiring left coronary artery images but they varies with different modalities and protocols especially when the volume of contrast matters in young infancy with heart failure.Diagnostic coronary angiography will provide more accurate,dynamic images and hemodynamic information in young infancy with ALCAPA and heart failure.

P2708 - EBSTEIN'S ANOMALY OF THE MITRAL VALVE REPORT OF A RARE CASE

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HCOR, PICU, São Paulo-Brazil[1]; H Santa Isabel, Pediatric Cardilogy, Salvador-Brazil[2]; HCOR, Radiology, São Paulo-Brazil[3]; HCOR, Echo, São Paulo-Brazil[4]; HCOR, Hemodinamic, São Paulo-Brazil[5] HCOR, Surgery, São Paulo-Brazil[6]

Case Report: S. A. B. A., 2 years old, with Interrupted Aortic Arch (IAA), Ventricular Septal Defect (VSD) and Mitral Valve Malformation, was submitted to pulmonary artery banding and stent placement in the arteriosus ductus in the neonatal period. One year later, she underwent a new procedure that included correction of the aortic arch and removal of both the pulmonary artery bandages. The correction of the mitral valve was not possible at the occasion. The best option was therefore to perform atriosseptectomy and pulmonary trunk banding. One year later, she evolved with significant ventricular dysfunction and then underwent nuclear magnetic resonance, which showed severe aortic valve stenosis and single papillary muscle in the inferior part of the mitral valve, with chordae tendineae for both valve leaflets, discrete insufficiency, and left ventricle dysfunction. In the echocardiogram, the valve showed two papillary muscles, with apical displacement of their leaflets and shortened chords, central coaptation in the apical region (Ebstein of the mitral valve), with discrete insufficiency and important stenosis. She was submitted to percutaneous aortic valve balloon valvuloplasty, which resulted in left ventricular function recovery. Five months later, she underwent mitral valve replacement with a mechanical prosthesis. The surgical description corroborates the reports of the imaging tests on the low implantation of the mitral valve and the subsequent atrialization of the LV.

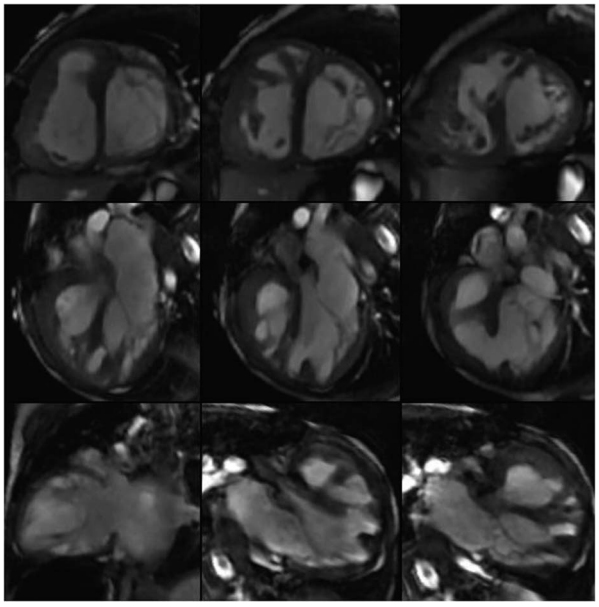

Figure 1.

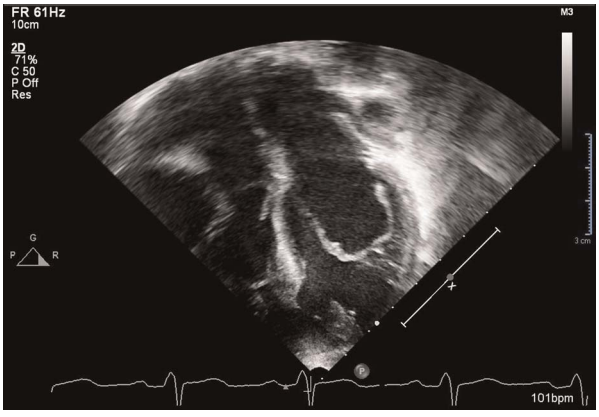

Figure 2.

P2732 - DOES PRENATAL DETECTION SCREENING INFLUENCE THE INCIDENCE OF CONGENITAL HEART DISEASE (CHD)

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Fello Pediatric Fellow Pediatric Cardiology, Cardiology, Bern-Switzerland[1]; Fellow Gynecology, Gynecology, Bern-Switzerland[2]; Staff Gynecology, Gynecology, Bern-Switzerland[3]; Head Pediatric Cardiology, Cardiology, Bern-Switzerland[4]; Head Pediatric Cardiac Surgery, Cardiology, Bern-Switzerland[5]; Staff Pediatric Cardiology, Cardiology, Bern-Switzerland[6]

Background: Detection rate for CHD in utero remains despite all efforts outside the perimeter of tertiary centers around 20-30% (FETCH data Switzerland 2005). To evaluate the screening success in the catchment area of the University Hospital in Bern we reviewed all in utero screenings from July 2009 to December 2014. In addition we looked at the incidence of CHD which should be around 0.8%. In Switzerland the current incidence of CHD is 0.5%.

Methods: Retrospective data analysis from July 2009 to December 2014 including all in utero and postnatal diagnosed new CHD at the Center of Perinatal Medicine at the University Hospital in Bern.

Results: 1522 in utero screenings have been performed. Of n = 339 patients with new CHD, n = 130 were born with an in utero diagnosis of CHD. n = 43 died in utero (detection rate of 51.03%). Median RACHS for prenatal diagnosed children was higher than in the postnatal detected CHD patients (2.4 vs. 3.1). The current incidence of CHD in the catchment area of Bern lays with 0.25% signficantly below the expected incidence of 0.6% in Switzerland.

Conclusion: This data clearly show that prenatal screening in the last years significantly improved from around 25% to currently 51% detection rate. In utero detected CHD has an overall higher RACHS in comparison with prenatally missed CHD, indicating that complex CHD is as of today better diagnosed in utero. Interestingly with a higher in utero detection rate we observe a decrease in the incidence of CHD postnatally.

P2752 - ANOMALOUS AORTIC ORIGIN OF CORONARY ARTERY. ¿AS FEARSOME AS WE THINK

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Background: Anomalous aortic origin of a coronary artery (AAOCA) is a congenital anomaly in which a major coronary artery arises from the wrong sinus of Valsalva and has an abnormal course. AAOCA is a cause of concern because its association with sudden cardiac death (SCD) during exercise. AAOCA with interarterial and intramural course are those with higher risk of SCD. Currently, there are no treatment guidelines for its management.
Methods: Review of patients with diagnosis of AAOCA, as an isolated malformation, in a Tertiary Hospital in the last 7 years.
Results: From 2009 to 2016, 5 AACAOS's cases were detected. The average age at diagnosis was 6.5 years (range 2-13 years). Most cases were detected in the study of a heart murmur. Only two patients presented symptoms (dyspnoea, chest pain and palpitations). In our study AAOLCA was more frequent than AAORCA (Table 1). None had interarterial nor intramural course. After the diagnostic suspicion by transthoracic echocardiography, the study was completed with ECG, Holter, Ergometry, Multi-cut CT scan (MDCT) and/or cardiac MRI. Suggestive alterations of ischemia were not detected. SPECT was performed in three patients and none showed defects of myocardial perfusion. Regarding treatment, all patients were advised to avoid exhausting excersice. Only two, who had symptoms at diagnosis, initiated beta-blocker treatment. Surgical treatment was not indicated in any patient. After a median of 2 years of follow-up, no patient presented a lethal event.
Conclusions: Patients with ACAOS who do not present interarterial or intramural courses should be considered of low risk and managed conservatively. After initial suspicion, performing a MDCT or MRI will allow to accurately delineate the coronary course and rule out the presence of high-risk anomalies. The presence of a normal effort test, especially associated with normal SPECT, will reaffirm us in the benign nature of this disease.

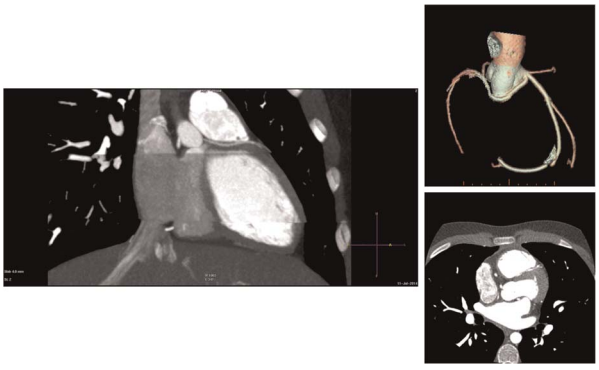

Figure 1.
Transeptal Anomalous aortic origin of left coronary artery: Single coronary arising from the right sinus (RCS). Anterior descending artery (ADA) with transeptal course and left circumflex artery (LCX) with retroaortic course.

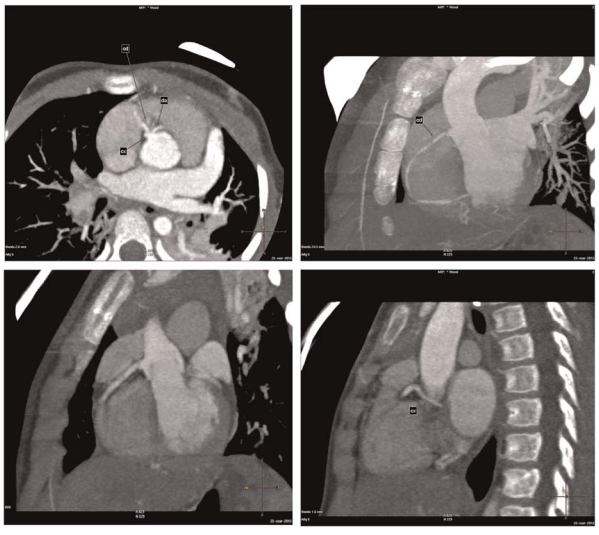

Figure 2.
Subpulmonary Anomalous aortic origin of left coronary artery: Single coronary arising from the right coronary sinus. Anterior descending artery with subpulmonary course and leftcircumflex artery with retroaortic course.

Table 1. Clinical features and management of Anomalous aortic origin of coronary artery's cases.

Sex Current age Age Diagnosis	Reasons for consultation at diagnosis	Type AAOCA
Female 12 years 10 years	Dyspnea and palpitations	**Transeptal AAOLCA** Single coronary arising from the right sinus (RCS). Anterior descending artery (ADA) with transeptal course and left circumflex artery (LCX) with retroaortic course.
Female 8 years 5 years	Cardiac murmur: Bicuspide aortic valve functionally normal	**Subpulmonary AAOLCA** Single coronary arising from the RCS. ADA with subpulmonary course and left circumflex artery (LCX) with retroaortic course.
Male 4 years 2 years	Cardiac murmur	**Transeptal AAOLCA** Single coronary arising from the RCS. Left coronary artery with transeptal course.
Male 15 years 13 years	Chest pain and dyspnea	**Retroartic AAOLCA** LCX hipoplastic arising from the RCS with retroaortic course. Right coronary artery and ADA normal origin and course.
Male 10 years 2,5 years	Cardiac murmur: Muscular VSD closed spontaneusly	**AAORCA** Single coronary arising from left coronary sinus giving origin to LCX, ADA and right coronary artery.

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Table 1. *Continued*

Sex Current age Age Diagnosis	Symptoms	Diagnostic Tests	Treatment
Female 12 years 10 years	Dyspnea, chest pain and palpitations during efforts	ECG: Normal Ergometry: Negative TTE: AAOLCA. Muscular apical VSD MDCT: AAOLCA SPECT: Normal	Atenolol Exhausting exercise was restricted
Female 8 years 5 years	Asymtomatic	ECG: Normal Ergometry: Negative TTE: AAOLCA. Bicuspid aortic valve. MDCT: AAOLCA SPECT:Normal	Exhausting exercise was restricted
Male 4 years 2 years	Asymtomatic	ECG: Nornal TTE: AAOLCA MDCT: AAOLCA	Exhausting exercise was restricted
Male 15 years 13 years	Dysnea and chest pain during efforts	ECG: Low T wave voltage of all leads. TTE: AAOLCA Ergometry: Not diagnostic. ST depression < 3mm (II, III, AVF) with chest pain. MDCT: AAOLCA SPECT: Normal	Atenolol Exhausting exercise was restricted
Male 10 years 2,5 years	Asymtomatic	ECG: Normal Ergometry: Negative Holter: Normal TTE: AAORCA MDCT: AAORCA CMRI: AAORCA	Exhausting exercise was restricted

**AAOCA: Anomalous aortic origin of coronary artery. AAOLCA Anomalous aortic origin of left coronary artery. AAORCA: Anomalous aortic origin of right coronary artery. VSD: Ventricular septal defect. ECG: 12-lead ECG. TTE: transthoracic echocardiogram. MDCT: Multi-cut CT scan. CMRI: Cardiovascular magnetic resonance imaging. SPECT: Myocardial perfusión SPECT.*

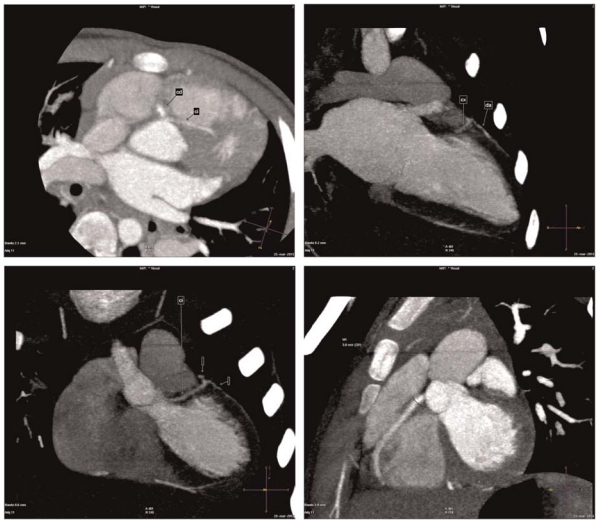

Figure 3.
Transeptal Anomalous aortic origin of left coronary artery: Single coronary arising from the right coronary sinus. Left coronary artery with transeptal course

P2754 - THE EVALUATION OF SYSTEMIC VENTRICULAR FUNCTION AFTER FONTAN OPERATION VIA CARDIOPULMONARY EXERCISE TEST AND TISSUE DOPPLER STUDY

Nimet Cindik[1], Birgul Varan[1], Kursat Tokel[1], Ilkay Erdogan[1], Mahmut Ozdemir[1], Murat Ozkan[2], Sait Aslamaci[2]
Baskent University, Pediatric Cardiology, Konya-Turkey[1]; Baskent University, Cardiovascular Surgery, Ankara-Turkey[2]

Background: Survival rates and quality of life of patients with single ventricle physiology were improved by Fontan operation. In our study, functional status and systemic ventricle function of patients with Fontan operation were evaluated.
Methods: 28 patients who underwent Fontan operation between January 1991 and December 2008 and who are capable of exercising were included. Cardiopulmonary exercise test, conventional echocardiography (ECHO), tissue Doppler Imaging (DTI), and PW Doppler ECHO were made and the relation between echocardiographic data, exercise capacity, and NT-proBNP levels were studied. 27 healthy children were the control group.
Results: The average age of operation was 5.6 ± 3.06 years, duration of follow up 6.85 ± 3.67, mean age 12.8 ± 4.36, mean age of control group 12.5 ± 3.76 years. There was a significant difference between patients and controls in terms of weight, height and BMI Z scores ($p < 0.05$). Cardiac output (CO), stroke volume (SV), systemic valve E velocity measured with PW, E/A ratio, systolic myocardial velocity (Sm), early diastolic myocardial velocity (Em), functional vital capacity (FVC), forced expiratory volume in 1 sec (FEV1), maximum voluntary ventilation (MVV), peak work, work at anaerobic threshold, peak heart rate, chronotropic index, peak oxygen consumption, peak carbondioxide production were all decreased in patients compared with control group ($p < 0.05$). Systemic ventricle myocardial performance index (MPI) calculated with DTI , septum MPI, MPI calculated with PW Doppler, dyssynchrony measurements, NT-proBNP levels were significantly higher in patients ($p < 0.05$).
Conclusion: Exercise test, NT-proBNP, dyssynchrony, and DTI measurements were found to be more sensitive than standard echocardiographic methods for the evaluation of clinical and functional status of the patients. Thus, serial measurements of NT-proBNP, dyssynchrony and DTI measurements of patients before operation and during follow up should be performed, cut-off values be determined and the relationship of the clinical and functional status with these parameters should be searched.

P2757 - FEASIBILITY AND REPRODUCIBILITY OF 3 DIMENSIONAL LEFT ATRIO VENTRICULAR VALVE SEGEMENTATION FROM TRANS THORACIC ECHOCARDIOGRAMS IN CHILDREN WITH CONGENITAL HEART DISEASE

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Boston Children's Hospital, Cardiac Surgery, Boston-United States[1]; Boston Children's Hospital, Cardiology, Boston-United States[2]; Children's Hospital, Cardiology, Boston-United States[3]; Children's Hospital, Cardiac Surgery, Boston-United States[4]

Background: Well developed algorithms/software exist for 3D segmentation of left atrioventricular valve (LAVV) using transesophageal echocardiography (TEE) in adults. However reproducibility of measurements is unknown in the pediatric population using transthoracic echocardiograms (TTE) particularly for precise annular diameter/annular angle measurements of the LAVV, that cannot be accurately determined from 2D images.

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Hypothesis: It is feasible, utilizing existing software, to semi-automatically segment annulus and leaflet of LAVV in pediatric population using 3D TTE with good reproducibility.
Materials and Methods: Full volume 3D acquisitions in the apical 4 chamber view obtained during routine clinical TTE imaging in 14 studies from 10 children, acquired using Philips IE33 with X3/5/7 probes, were segmented 3 times each by 2 independent reviewers using the 4D mitral valve assessment package (TomTec 3.1). Maximum differences in the measurements between and within observers was determined. Error rates were calculated as follows: (maximum difference/mean)×100.
Results: Pathologic and normal mitral valves, in children aged 0.2 to 5.3 years, weight 4.4 to 29.3 kg were segmented. AP diameter, AL-PM diameter, sphericity index and nonplanar angles were derived. Error rates between and within observers are depicted in the figure and varied by quality of image, with smaller variation in higher quality imaging. The average error rates ranged between 10-18% for measurements of interest between observers with lower error rates within observers.
Conclusions: High quality transthoracic images of the LAVV can be reproducibly segmented using current commercial software, enabling better analysis of diseased LAVV in small children in whom 3D TEE is not applicable.

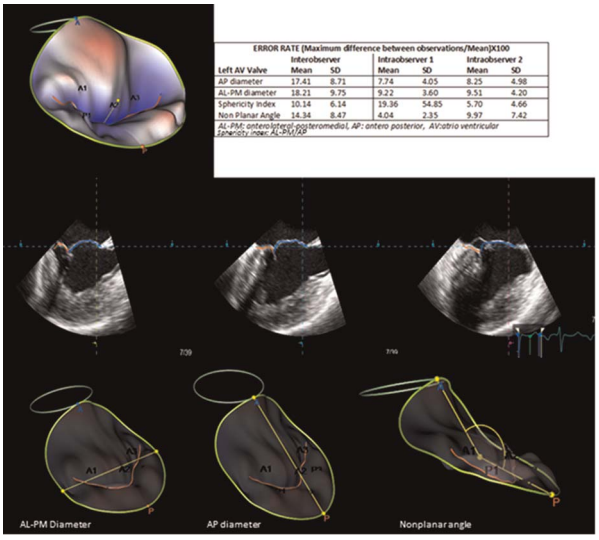

Figure.

P2761 - EXERCISE PERFORMANCE ASSESSED BY CPET IN FONTAN CHILDREN AND YOUNG ADULTS

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Background: Exercise capacity(ExC) after the Fontan procedure (FP) is reduced. The following mechanisms explain why patients with Fontan physiology have a restricted ExC (reduced peak oxygen consumption, VO2): failure to increase stroke volumen, reduced preload(lack of subpulmonary pump), cronic hipoxemia, chronotropic impairment, musculoskeletal deconditioning(functional deficiency of the peripheric muscle pump).
Objectives: To define exercise performance quantitatively through cardio pulmonary exercise test (CPET) in children and young adults after FP.
Methods: Retrospective observational study of 72 children and young adults (56% male, average weight 41.9 ± 17 kg, average height 146.5 ± 17.5 cm) aged between 6-19 years (11.8 ± 3.4) who have undergone a FP. Ramp treadmill (Bruce protocol) was performed. Data were analyzed for population that achieved a maximal aerobic capacity. Inclusion criteria admited patients with either right or left systemic ventricles. 9 subjects with pacemakers, sinus node dysfunction or junctional rhythm were excluded.
Results: Submaximal exercise tests limited by symptoms were performed, clinically and electrically negative in all cases, with no significant arrhythmias. Effort time 10.22 ± 2.4 min (8.8 ± 2.4METs). Rest HR 95 ± 17bpm, maximal HR 164 ± 26bpm (HRR 69 ± 27bpm). 24 patients reached 85% predicted HR (mean 77 ± 13.5%). SBP/DBP at rest was 102 ± 14/59 ± 9 mmHg, maximum of 127 ± 19/69.5 ± 9 mmHg. DP was 20950 ± 5572.2. 39 patients had normal spirometric pattern,18 restrictive. Exercise function of these patients varied greatly. In some, exercise capacity was well preserved, with normal and even above-average peak VO2 whilst in other cases peak exercise capacity was significantly depressed. CPET showed decreased maxi VO2 (76 ± 19.8% predicted), with normal O2 pulse (102.16 ± 33% predicted, curves with early rise and maintained plateau). 39 patients did not reach MPHR, wich may suggest chronotropic impairment. The rise in Vslope and CO2Eq at VAT (38 ± 6.8 and 39.3 ± 7.5 respectively) translated ventilatory inefficiency(V/Q imbalance, according to Fontan physiology), with an appropriate respiratory reserve.
Conclusions: They remain limited in their physical abilities with maximum VO2 around 76% of predicted value compared to their peers in our sample. Decreased ventilatory efficiency its characteristical. CPET is an useful tool to assess ExC after the FP, providing relevant data: although cardiac otuput depends greatly on preload for these patients, in a sample without intrinsic limitations of the systemic ventricle chronotropic response is imperative to obtain a suitable VO2.

P2765 - REALIZING AUTOMATED ECHOCARDIOGRAPHY FOR NEWBORNS AND INFANTS

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Despite progress in reducing the size and cost of pediatric echocardiography, significant gaps remain in the ability to obtain quality pediatric heart and lung imaging for the youngest patients. Following a modest research grant from the from the Pediatric Device Innovation Consortium via the University of Minnesota's Office of Discovery and Translation, the Newborn Foundation and its program investigators have worked to develop a proof of concept for an automated array of transducers to provide simplified, quick and standardized acquisition of diagnostic-quality echo images to be obtained by individuals with little or no technical training or expertise. The aim is to facilitate highly automated image acquisition to improve access and outcomes with faster,

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remote interpretation. The presentation will address the current stage of proof of concept software and hardware, along with the landscape of CHD and RHD requiring echo imaging for the youngest patients – and how such technology could improve care and outcomes for these patients. Images obtained can be shared with specialists or potentially evaluated with on-board machine learning technologies. Such a device has applications in the developed and developing world. Access to confirmatory pediatric echo for babies failing newborn CCHD screening is still difficult to obtain in many parts of the United States, while also providing a mechanism to better guide remote care and referrals for young children with suspected or confirmed heart disease that have no local access to pediatric cardiology specialists. In the developing world, the signs and symptoms of both newborn infections and pediatric cardiac abnormalities can be difficult for health workers to detect. This device would support an increased body of literature on the value of ultrasound in diagnosing both CHD and childhood pneumonia with greater sensitivity and specificity than existing methods, including x-ray/radiation.

P2767 - CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES IN THE FETUS. A RETROSPECTIVE ANALYSIS OF ECHOCARDIOGRAPHIC FEATURES AND OUTCOME IN A TWO CENTER SERIES OF 30 CASES

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Objectives: to describe the echocardiographic features and outcome of fetuses with congenitally corrected transposition of the great arteries (ccTGA).
Methods: 30 consecutive fetuses were diagnosed with ccTGA at one of two referral centers between 2005 and 2015.
Results: median gestational age at diagnosis and at last fetal echocardiogram were respectively 23 and 35 weeks. Most fetuses were referred for suspicion of cardiac malformation. Only 2/30 (7%) fetuses presented an isolated form of ccTGA. Associated cardiac defects included: abnormal cardiac position in 11/30, ventricular septal defect (VSD) in 25/30, pulmonary atresia in 8/30 and pulmonary stenosis in 6/30, abnormal tricuspid valve in 7/30, right aortic arch in 2/30, suspicion of aortic coarctation in 4/30. Two fetuses developed a complete atrioventricular (AV) heart block in the third trimester and required a permanent pacemaker implantation at birth. Only one fetus presented an extracardiac anomaly. Karyotype was tested prenatally in 11/30 fetuses and resulted normal. Six pregnancies were interrupted and 2 fetuses were lost at follow up. Mean gestational age at birth was 39 weeks, mean birth-weight 3200 g and mean oxygen saturation 95%. 17/22 patients required one or more surgical procedures. Among them, 2 completed the Fontan circulation, 2 received a bidirectional cavopulmonary connection, 5 the anatomic repair and 2 the physiologic repair. At a mean follow up of 5,2 years (±3,5) 4/22 patients were lost at follow up whereas the others are alive and clinically well.
Conclusions: isolated cases of ccTGA and associated extracardiac or chromosomal anomalies were rare prenatally. The mid term outcome was reasonably good for all liveborn infants even if most of them needed one or more cardiac treatment. Prenatal counselling remains challenging because also fetuses with an apparently favorable condition may develop major changes particularly regarding to tricuspid valve and cardiac rhythm with significant changes in prognosis.

P2771 - PULMONARY PERFUSION IN PATIENTS WITH HYPOPLASTIC LEFT HEART SYNDROME AFTER FONTAN OPERATION

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Introduction: Fontan operation results in diminished pulmonary blood flow pulsatility, affecting pulmonary blood distribution. Left pulmonary artery (LPA) stenosis is a common complication of multistage HLHS palliation. Aim: Assessment of lung perfusion in HLHS patients after Fontan completion using planar lung perfusion scintigraphy.
Material and Methods: 77 patients (55 boys – 71%), mean age 9.3 years ± 3.2 underwent planar lung perfusion scintigraphy: two injections of 99mTc – MAA into the right upper, then into the right lower extremity. Webster's formula was used to calculate the activity and Hawkeye hybrid gamma camera to perform lung perfusion studies. Percentage share of every lobe and lung in a global lung perfusion was calculated after averaging of counts in anterior and posterior projection (geometric mean) for inflow from superior and inferior cavopulmonary anastomosis. 58 patients (75%) had LPA stenting.
Results: Hypoperfusion (lung contribution to global lung perfusion $\leq 35\%$) of the left lung in 40,3% (31/77). Perfusion of the lungs and lobes wasn t significantly higher in patients with stent ($n = 58$). Lowest perfusion - upper lobe of the right lung. It was significantly lower than in the middle and lower lobes of the same lung and the upper and lower lobes of the left lung. ($p < 0.0001$). Perfusion of the left lung and respective lobes wasn't significantly higher in patients with stent ($n = 58$). Frequency of hypoperfusion of the left lung wasn't significantly lower in patients with stents than without: 37.9% (22/58) vs. 47.4% (9/19). ($p = 0.47$). Postoperative diaphragmatic paralysis was noted in 5 patients (6,5%).
Conclusions: 1. Non-physiological pulmonary blood flow, pulmonary artery obstruction and postoperative diaphragmatic paralysis contribute to abnormal pulmonary perfusion in patients with HLHS. 2. Hypoperfusion of the left lung is the most common finding despite successful LPA stenting. 3. Planar lung perfusion scintigraphy is a reliable tool in evaluation of patients after Fontan completion.

P2773 - EVALUATION OF THE VENTRICULAR FUNCTIONS ASCENDING AORTA AND AORTIC ROOT WIDTHS IN THE ASYMPTOMATIC PATIENTS WITH ISOLATED BICUSPID AORTIC VALVE

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Objective: Aortic root and/or ascending aorta dilatation is frequently encountered in patients with bicuspid aortic valve(BAV). The aim of this study is to evaluate the ventricular functions, ascending aorta and aortic root widths in asymptomatic patients with isolated BAV to define their prognosis.
Method: 3 to17 years old (mean 9,5 ± 3,9) 49 patients (41 males, 8 females) with BAV with/out stenosis or insufficiency and 3 to

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15 years old (mean 8,8 ± 2,9) 20 healthy controls (15 males,5 females) were enrolled in this study. Their aortic root, ascending and descending aorta widths, aorta and sinus valsalva elasticities were measured. Aortic valve morphology, valve regurgitation(AR) jet length and directions were recorded. Tissue Doppler Imaging(TDI) was performed to obtain views from interventricular septum(IVS), mitral and tricuspid free wall in apical four chamber position. Blood natriuretic peptide(BNP) levels were measured.
Results: No difference was found between the age, height, weight, systolic and diastolic blood pressures of two groups. The most common valve morphologies in these patients were: right to left cusp fusion (40.8%), right non-cusp fusion (36.7%), bicuspid (18.3%) and left non-coronary cusp fusion (4.08%). The most common two directions of AR jets were central (27%) and left eccentric (26%). Neither stenosis nor insufficiency was observed in 11(22.44%) patients. Ascending aorta systolic (2.29 ± 0.36 and 2.06 ± 0.29 cm, $p < 0.05$) and diastolic (2.08 ± 0.36 and 1.90 ± 0.27 cm, $p < 0.05$) measurements of the patient group were found significantly higher than the control group. Mitral valve early and late diastolic (MA) flow velocities of the patient group were significantly lower than the controls (E:0.97 ± 0.13 A = 0.59 ± 0.13, E:0.89 ± 0.13 A: 0.50 ± 0.07 m/sn). IVS early diastolic flow velocity of the patient group was significanly lower than the controls in TDI (11.28 ±1.82 and 12.9 ± 1.73, $p < 0.05$). A positive correlation was found between BNP and ascending aorta stiffness, ascending aorta systolic, descending aorta systolic and diastolic diameters, also betweenAR length.

P2775 - RADIATION DOSE IN THE HYBRID OPERATING ROOM USED FOR PEDIATRIC CARDIAC SURGERY

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Nowadays we can observe an increasing number of hybrid procedures in treatment of complex congenital heart defects (CHD). The radiation dose of hybrid procedures is still underrated. There are no guidelines determining radiation dose levels for pediatrics hybrid procedures. The main purpose of this study was to assess the radiation dose during hybrid procedures in CHD correction. We retrospectively analyzed 81 patients who had had hybrid CHD surgery in a hybrid operating room (Siemens Axiom Luminous). Fifty-seven (70.37%) had been subjected to CHD correction without stent implantation, 10 patients (12.34%) had undergone pulmonary arterioplasty; six (7.4%) - stent implantation; three (3.7%) heart valvuloplasty, and five (6.17%) - ECMO implantation. Boys predominated (65.43%). Forty-four patients (54.32%) were treated during the first year of life, the median weight was 7.4 kg. 7778% of the patients had combined CHD, at least 70% of them had Hypoplastic Left Heart Syndrome (HLHS).The median fluoroscopy time for all the procedures was 3.8 min. The median Skin Entrance Dose (SED) for all the procedures was 38 mGy; Air Kerma (K)-74.3 mGy and Dose Area Product (DAP) 516.34 μGym2. The median exposure time for HLHS treatment was 1.9 min. for the Norwood (SED = 31; DAP = 270.06; K = 63.5) procedure; 2.8 min. for hemi-Fontan (SED = 74; DAP 447.65; K = 71.7) and 3.8 min. for Fontan (SED = 211; DAP 1178.3; K = 456). Hybrid procedures for CHD treatment appear to be a radiation-safe method. The mean radiation doses are much lower than doses used in computed tomography scans and comparable to doses received during several chest X-ray procedures. Radiation monitoring is an important component of pediatric cardiac surgery. Further studies are needed to evaluate the radiation levels in hybrid pediatric cardiac surgery.

P2780 - EPIDEMIOLOGY CORONARY ARTERY LESIONS AND TREATMENT OF KAWASAKI DISEASE IN CHILDREN FROM ARGENTINA PERIOD JUNE 2014 TO DECEMBER 2016

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Background: Although Kawasaki disease (KD) is the leading cause of pediatric acquired heart disease in many countries, there are few indexed published studies from Latin America (LA). We describe the epidemiology, coronary findings and treatment of children with KD at two centers as part of the prospective REKAMLATINA-1 Network surveillance study of KD in LA.
Methods: Prospective descriptive study of patients (pts) with a hospital discharge diagnosis of KD according to AHA criteria, who were admitted at 2 hospitals in Argentina. Study period: June-1-2014 to Dec-31-2016
Results: 19 pts were analyzed, 11 (57.9%) were male. All were hospitalized; mean length of hospitalization was 5.5 (3-12) days. Median age was 45 (7-98) months. Distribution by age groups was: <6 months, 0 (0%) pts; 7-24 m, 3 (15.8%) pts; 25-60 m, 12 (63.2%), and >60 m, 4 (21.0%) pts. Mean duration of fever at admission was 5.5 (1-12) days. Prior to KD final diagnosis, 9 pts (47.4%) received antibiotics for other presumed diagnoses. Baseline echocardiogram (ECHO) was performed in all pts, of which 6 (31.6%) pts had >1 abnormality documented, including coronary artery lesions (CALs) in 4 (21.1%) pts: dilation (3 pts) and giant coronary aneurysms (1 pt). A second follow-up ECHO was performed in all 19 pts, of which CALs were documented in 1 (5.3%) pt. All 19 pts received IVIG: 1 dose, 18 (94.7%), and 2 doses, 1 (5.3%), respectively. Among those pts treated with IVIG at admission, 1 (5.3%) had >10 days of fever. Aspirin was given in 100% and none received steroids. No deaths occurred.
Conclusions: The percentage of CALs in this analyzed group is higher than similar patients from other LA centers who were adequately treated with IVIG and aspirin.

P2784 - MANAGEMENT AND OUTCOMES IN BORDERLINE LEFT VENTRICLE A SINGLE CENTER 6 YEAR EXPERIENCE

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Background: Patients with borderline left ventricle represent, the most challenging group when determining a surgical management strategy since no scoring system reliably predict long-term outcomes. We assess echocardiographic characteristics, surgical management and outcomes of this patients.

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Materials and Methods: Retrospective descriptive analysis of patients with borderline left heart presenting between January 2010-April 2016. Echocardiographic features were reviewed, Rhodes's, Collan's and UVR-SA scores were applied. The outcome regarding the surgical election, recruitment procedures and the need for reintervention at follow-up were evaluated.
Results: A total of 26 patients were included, 65% of them were male. One patient died before surgery because of comorbidities (Edward's syndrome and prematurity)and was excluded. Biventricular repair surgery was performed in 23 patients (92%). Of those, two patients died due to failed bi-ventricular repair and one died due to hemochromatosis. Recruitment procedures were performed in 92% of biventricular repair. The need for reintervention was 52% and all had a good functional class at 6 years of follow-up. Two (8%)patients had univentricular repair which initially hybrid procedure due to prematurity. Both patients died. Rodhes's score classified correctly 7 out of 25, Collan's 13 and UVR-SA 4.
Conclusions: From our serie,we conclude that echocardiographic scores don't predict long-term outcomes in patients with borderline left ventricle, probably because of the high percentage of recruitment procedures that allowed a successful biventricular surgery. In the 2 cases of failed biventricular repair, the three analysed scores would have predicted a greater survival with single ventricle palliation. Nowadays survival with univentricular surgery improved meaningfully, but long-term results are already poor. Our patients were a high risk group with a lot of comorbidities. We observed low mortality (4.5%) with biventricular surgery despite the high need for reintervention. We speculate that biventricular physiology should be preserved with accurate patient selection. We believe the echocardiographic score systems should be revised to include left ventricle recruitment strategies in order to accurately predict overall surgical management and long term outcomes of patients with borderline left ventricle.

Table 1.

		Rodhes	Collan (with EFE)	Collan (without EFE,	UVR-SA			Rodhes	Collan
BVR	Correctly classified	3	9	6	6	BVR	PPV	100%	100%
	Incorrectly classified	0	0	0	0		PNV	18%	25%
UVR	Correctly classified	4	4	4	4	UVR	PPV	18%	25%
	Incorrect classified	18	12	15	21		PNV	100%	100%

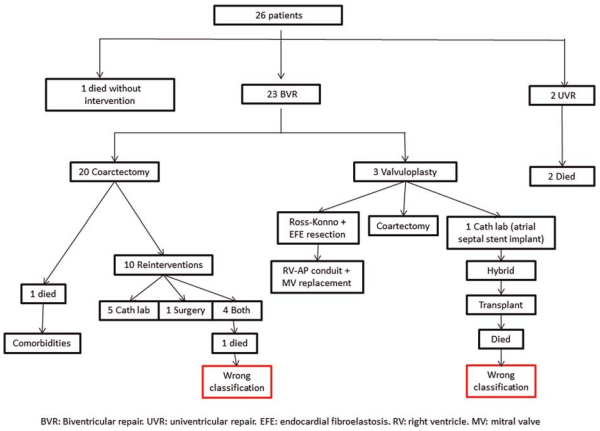

Figure.

P2792 - COMPLEX ANOMALY OF AORTIC ARCH IN PHACE SYNDROME A CASE REPORT

Ahmet Vedat Kavurt[1], Feyza Ayşenur Paç, Deniz Eriş, Ibrahim Ece[1], Serhat Koca[1], Mustafa Özdemir[2]
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İntraPHACE syndrome (posterior fossa anomalies, hemangioma, arterial lesions, cardiac abnormalities/coarctation of the aorta, eye anomalies) is characterized by large infantile hemangiomas (IH) of the face, neck, and/or scalp that are associated with developmental defects. The case of a 5-month-old male infant with PHACES syndrome is reported. On physical examination the infant had infantile hemangioma (6 × 7 cm) extending from the left temporal region of the scalp to his neck. Frontal bossing, plagiocephaly, micrognathia, sternal pit and tag were noted . Cardiac computed tomographic angiography (3D-CTA) and catheter angiography revealed left-sided cervical aortic arch, aortic arch dysplasia (kinking, looping, tortuosity, aneurysma , hypoplasia), coarctation of the aorta (at the level transverse arch) and aberrant left subclavian artery originates from a proximal descending aorta. Segmental absence of left internal carotid artery (ICA) and

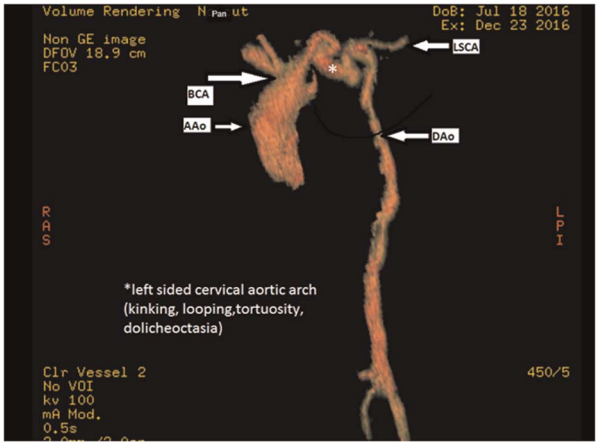

Figure 1.

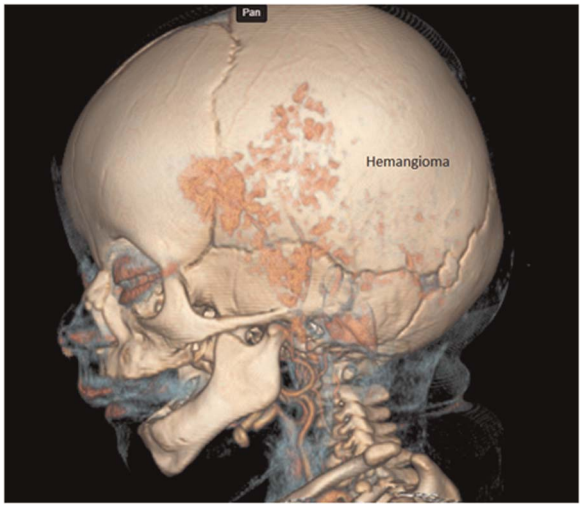

Figure 2.

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moya-moya like collateral arteries, absence of right posterior communicating artery were detected on the 3D cranial CTA. Ophthalmological examination was normal. Cranial magnetic resonance imaging (MRI) showed no cerebral and posterior fossa anomaly. In conclusion, PHACE is associated with a high risk of congenital heart disease and cerebral arterial anomalies. Cardiac and aortic arch imaging with detailed assessment of arch patency and brachiocephalic origins is essential for any patient suspected of having PHACE.

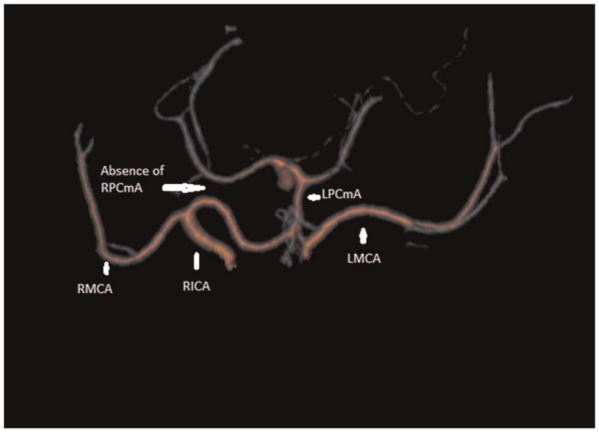

Figure 3.

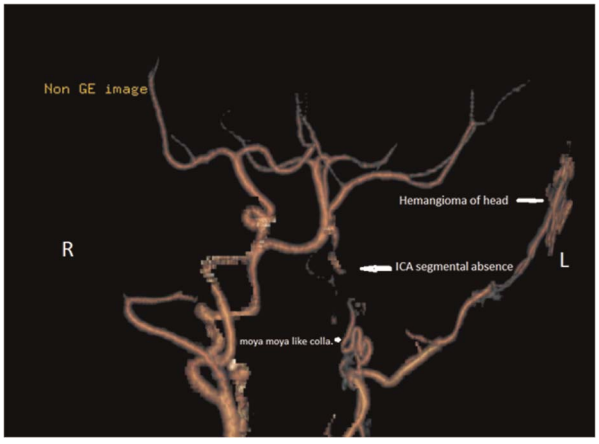

Figure 4.

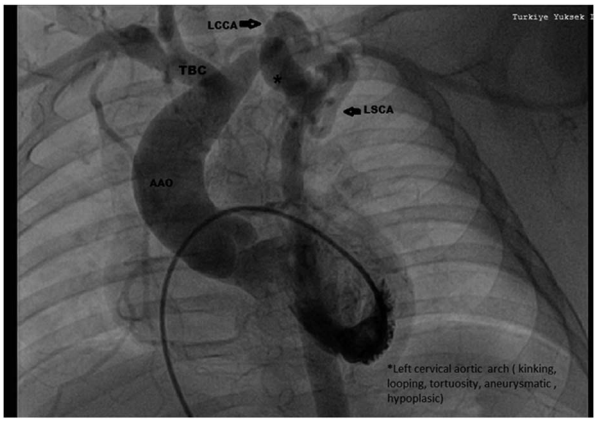

Figure 5.

P2801 - RV MYOPATHY IN CHILDREN CARDIAC MAGNETIC RESONANCE FEATURES

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Background: Right ventricular cardiomyopathy (RVCMP) in children and adolescents is a poorly described entity. Conventional transthoracic echocardiogram is a suboptimal tool to evaluate the right ventricle. But, cardiac magnetic resonance imaging (CMR) is emerging as the modality of choice for comprehensive evaluation, including tissue characterization in cardiomyopathies. We present CMR features of 5 children with cardiomyopathy predominately involving the right ventricle.
Material and Methods: Our study group consists of children aged 7months – 15 yrs, weighing 4 kgs - 53 kgs, with 4 boys and 3 girls. All presented with features of right heart failure without arrhythmia. All appropriate seqences were acquired. Gadolinium was used as contrast agent. Late gadolinium enhancement (LGE) images were obtained to look for myocardial fibrosis.
Results: All 7 children had significantly dilated, IVC , RA and RV. The body surface area Indexed RV EDV ranged from 120 ml/M2 to 395 ml/M2. The RVEF varied from 11% to 38%. Four children had exclusive RV failure with normal LV systolic function. In 2 other children there was more RV failure than LV (RVEF 20% vs LVEF of 35%, 23% vs 31%). All had siginificant TR, one had severe PR and another moderate MR. 2 had preserved RV apical function, distal to the moderator band while the rest had global hypokinesia. LGE imaging showed, patchy enhancement in 4. 3 children showed no LGE, which may suggest an absence of RV myocardium as biopsy suggestive of uhl's anomaly.
Conclusions: RVCMP in childhood is a rare entity and the aetiology is not clearly understood. Whether it is a childhood form of the better-described ARVD in the adults is unclear. CMR is a very useful comprehensive tool for evaluation.

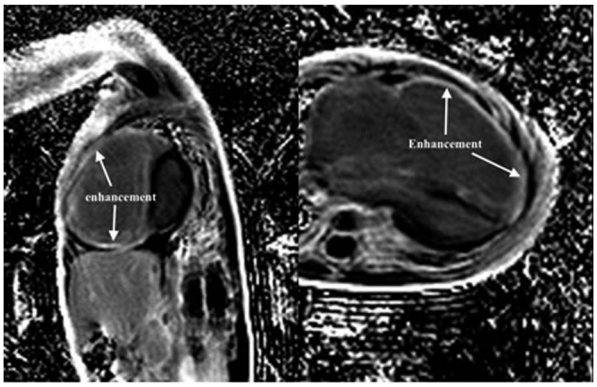

Figure.

P2810 - SPONTANEOUSLY REGRESSION OF THE RIGHT VENTRICULAR DIVERTICULUM AND VENTRICULAR SEPTAL ANEURISM

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Background: Right ventricular diverticulum (RVD) and ventricular septal aneurysm (VSA) are rare congenital cardiac malformations.

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RVD are scarcely diagnosed during the fetal period. To our knowledge, this is the first case that VSA associated with RVD. In the present study, we report a case of RVD associated with VSA and its spontan regression.
Case Report: A 34 year old multigravida was referred to our ultrasound unit at 22 weeks gestation for suspected echogenic focus. The four chambers view of the fetal heart revealed an aneurysmatic enlargement was observed in the right ventricular lateral-apical wall. This aneurysmatic enlargement is separated from the right ventricle by an area of 4 millimeters. Color Doppler detected blood flowing into and out from this sac, suggesting a right ventricular diverticulum. Echocardiograpy performed every 4 weeks during pregnancy did not detect any progression in the size of the diverticulum. A female infant was born spontaneously at 39 weeks. In addition RVD, ventricular septal aneurysm was detected by postnatal echocardiograpy and cardiac-MRI (figure 1,2, white arrow: RVD, black arrow: VSA). Conservative management was planned. He was followed by echocardiography and electrocardiography at intervals of 10 days. RVD and VSA were spontaneously regression. It was observed that RVD and VSA were fully recovered at the second month control.
Conclusion: We present a case of prenatal diagnosis of right ventricular diverticulum at 22 weeks gestation. In addition RVD, VSA was detected by postnatal echocardiography. The diverticulum and VSA disappeared before the age of 2 months. Echocardiography of the patient at 5 months of age revealed no cardiac diverticulum and aneurism or any cardiac anomaly and showed normal heart function.

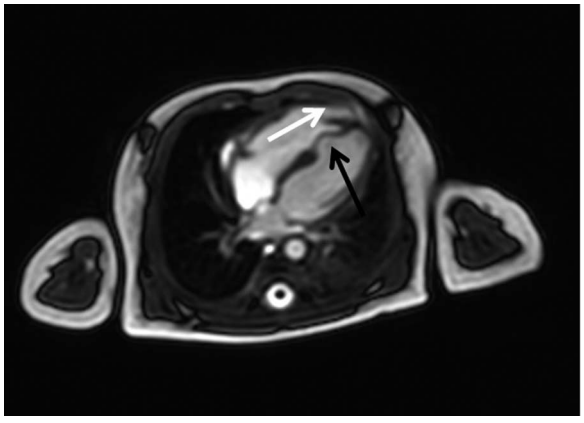

Figure 1.

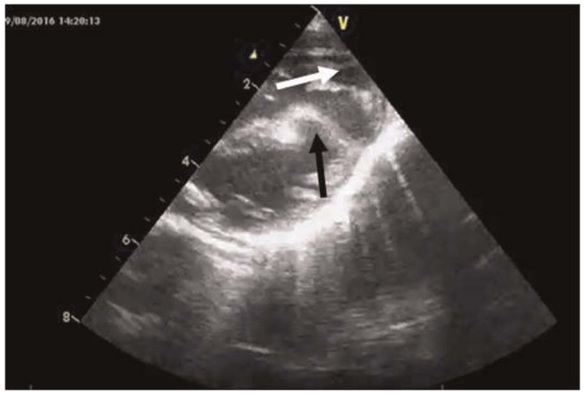

Figure 2.

P2818 - CORRELATION OF ECHOCARDIOGRAPHIC MEASUREMENTS TO EXERCISE PARAMETERS IN FONTAN PATIENTS A PROSPECTIVE BLINDED STUDY

Suma Goudar, Daniel Forsha, David White, Mark Gelatt, Tyler Johnson, Rita France, William Chew, Girish Shirali
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Background/Hypothesis: While many echocardiographic (echo) parameters are used to quantify ventricular function, it is unclear which measurements correlate with functional capacity in patients with single ventricle physiology. The purpose of this study is to compare quantitative echo measurements of ventricular function to measurements of functional capacity by exercise stress test (EST) in single ventricle children post-Fontan completion.
Materials and Methods: EST was performed via cycle ergometer (James protocol) to measure peak oxygen consumption (VO2peak) and VE/VCO2 slope in 28 Fontan subjects from December 2015-January 2017. Resting echocardiograms were performed on the same day as EST using GE Vivid9. Separate, blinded observers performed all Echo and EST measures. Global longitudinal strain (GLS) calculated via speckle-tracking analysis using EchoPAC. Qualitative atrioventricular valve regurgitation (AVVR) and ventricular function was graded an ordinal scale, with the highest score equal to worsened AVVR. Correlation was performed using Pearson Correlation. Multivariable regression model was used to determine interaction between variables with significant correlation.
Results: 28 subjects aged 10-17 years were analyzed. VO2peak ranged from 13-31.06 ml/kg/min and global longitudinal strain (GLS) ranged from -7.4 to –22.6. GLS and AVVR were the only two echo variables with significant correlation with VO2peak (-0.45 and -0.46 respectively, $p = 0.01$). There is no significant interaction between GLS and AVVR ($p = 0.11$) and each variable is an independent predictor of VO2 max on multivariate regression. All other echo variables (notably E/A, E/E', S', fractional shortening, fractional area change, and 2D ejection fraction and AV valve annular plane systolic excursion) had poor correlation with peak VO2. No echo variables correlated with VE/VCO2 slope.
Conclusions: In this cohort, GLS and AVVR severity were the only echo measures associated with peak VO2 by exercise testing. These findings should be considered in designing protocols for quantification of ventricular function by echo post Fontan.

P2836 - POSTOPERATED TETRALOGY OF FALLOT AND FONTAN PROCEDURE CHILDREN WHY SIMILAR CPET RESULTS

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Introduction: Postoperated children with complex congenital heart disease (CHD) have a decreased functional capacity (FxC) associated with death and hospitalization. Cardio pulmonary exercise test (CPET) assesses their FxC and different specific patterns are expected, according to different physiological adaptations depending on the type of CHD.
Objective: The aim of this investigation was to compare CPET between postoperated Tetralogy of Fallot children (PTF) and subjects that had undergone a Fontan procedure (FP).

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Methods: Retrospective observational study of 25 PTF (mean age 12 ± 3.2 years, weight 43.2 ± 14.55 kg, height 146.7 ± 14.4 cm,58% male) and 63 Fontan (mean age 11.8 ± 3.4years, weight 41.9 ± 17 kg,height 146.5 ± 17.5 cm, 56% male). Inclusion criteria admited pulmonary atresia with VSD in the PTF group and both right or left systemic ventricles in the FT group.Patients with pacemakers, sinus node dysfunction or junctional rhythm were excluded. Ramp treadmill ergometry (Bruce protocol) was performed with expired gas in all cases.
Results: Submaximal exercise tests limited by symptoms were performed, clinically and electrically negative in all cases,with no significant arrhythmias. We found no significant differences between both groups ($p > 0.05$) despite some comparisons were remarkable: 38% of FP reached 85% MPHR vs 32% in PTF, in FP group 62% showed a normal spirometric pattern and 28.6% restrictive vs 56% with normal pattern and 20% restrictive in PTF group. CPET results were:(image1) From a cardiovascular stadpoint VO_2 was decreased in both groups, revealing impaired ExC with normal O_2 pulse (appropriate stroke volume). FP group showed ventilatory inefficiency with higher V slope and CO_2 equivalents (V/Q disturbance), whilst in the PTF group those parameters were limit.
Conclusions: FxC is decreased in both PTF and FP patients. We found no significant differences beetwen them that may be attributable to a bigger sample of FP and their heterogenicity.Ventilatory efficiency variables revealed V/Q imbalance for FP subjects (according to their condition). This disturbance is only appreciated in PF when significant RV dysfunction develops.

Table.

	FONTAN procedure N = 63	Postoperated T. of FALLOT N = 25
Effort time (min)	10.22 ± 2.4	10.3 ± 2.7
METs	8.8 ± 2.4	9.3 ± 3
Resting HR (lpm)	95 ± 17	89 ± 13.8
Max HR	164 ± 26	169.7 ± 17.3
HRR	69 ± 27	80.7 ± 22.3
Max predicted HR (%)	77 ± 13.5	79.6 ± 9.3
Resting SBP/DBP (mmHg)	102 ± 14/59 ± 9	97.4 ± 10/56.6 ± 8.4
Max SBP/DBP (mmHg)	127 ± 19/69.5 ± 9	125.4 ± 10.3/71.2 ± 7.5
Double product	20950 ± 3372.2	20938 ± 2817
Max VO_2 (% predicted)	76 ± 19.8	79.5 ± 10.2
O_2 pulse(% predicted)	102.16 ± 33	101 ± 18.5
VE/VCO_2 slope (°)	38 ± 6.8	33.4 ± 8.3
O_2 equivalents	36.9 ± 6.2	32.7 ± 7.3
CO_2 equivalents	39.3 ± 7.5	34.9 ± 6.8
Respiratory reserve	39 ± 12.3	42 ± 14.6
OUES	1.47 ± 0.55	1.57 ± 0.51

P2849 - SINGLE CASE OF RIGHT CORONARY ARTERY ANOMALY IN INFANCY IN 15 YEAR STUDY IN OUR TERTIARY CARDIOLOGY CENTRE

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Background: Anomalous right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital defect affecting 0.002% of the population. Unlike anomalous left coronary artery from the pulmonary artery (ALCAPA), ARCAPA generally has not been considered to be a lethal defect in infancy. High risk group for sudden cardiac death is common after vigorous exertion in children and teenagers, and majority are asymptomatic without previous complaints of cardiac symptoms (palpitations, chest pains, collapse).
Methods: We describe a case of 1 yr old asymptomatic boy, who was referred from local hospital for evaluation of a heart murmur. Patient had echocardiography study and CT angiography of coronary arteries which demonstrated an anomalous right coronary artery from the pulmonary artery with dilatation of the left coronary artery and numerous tortuous left coronary artery collaterals. Myocardial strain and tissue Doppler showed preserved function of the right ventricle. The management decision was to proceed with elective surgical correction of right coronary artery within 3 months post diagnosis.
Conclusion: Anomalous right coronary artery from pulmonary artery is a rare congenital coronary abnormality. We described for the first time right ventricle deformation in an asymptomatic patient.

P2852 - MYOCARDIAL EXTRACELLULAR VOLUME FRACTION QUANTIFIED BY CARDIAC MAGNETIC RESONANCE IN CHILDREN

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Background: Disturbances in the myocardial extracellular volume fraction (ECV), such as diffuse or focal myocardial fibrosis or oedema, are hallmarks of heart disease. Diffuse ECV changes are difficult to assess or quantify with cardiovascular magnetic resonance (CMR) using conventional late gadolinium enhancement (LGE), or pre- or post-contrast T1-mapping alone. ECV measurement circumvents factors that confound T1-weighted images or T1-maps, and has been shown to correlate well with diffuse myocardial fibrosis. The aims were to assess paediatric data for a variety of clinical conditions and to detect sub-clinical abnormalities in 'normal appearing' ventricular function.
Methods: ECV maps were generated from T1-maps acquired pre- and post-contrast calibrated by blood hematocrit. As a part of the CMR study (between March-December 2016), they underwent a pre- & pos-contrast acquisition of a single breath-hold Modified Look-Locker Inversion-recovery (MOLLI) pulse sequence in a single mid-ventricular short axis imaging plane.
Results: 20 paediatric patients (mean age 12.2 years, range: 5-18 years) were assessed with full CMR study. We looked at all the 6 segments (based on conventional 17-segment model) and their ECV values from 0.3 to 0.4, with incremental of 0.01 to see for the number of segments affected and compared this to the normal adult data and also with the global ejection fraction. As expected, those with lower ejection fraction tended to have more affected segments, but we showed that even those with normal function had abnormal segments.
Conclusions: Extracellular volume fraction imaging can quantitatively characterise segmental changes, and subtle myocardial abnormalities not clinically apparent on global LV function & LGE images. Taken within the context of adult literature, and paucity of paediatric data, more work needs to be done to develop this as a clinical tool within paediatric cardiology.

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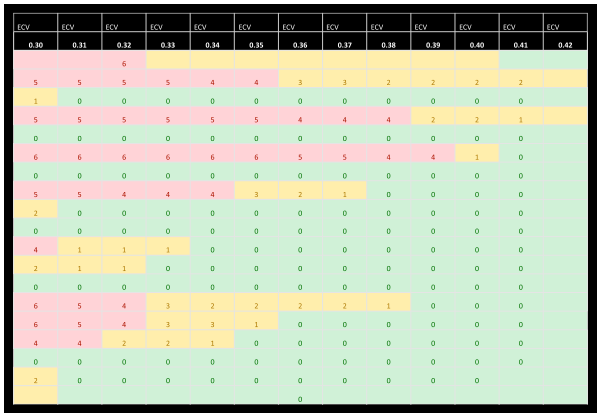
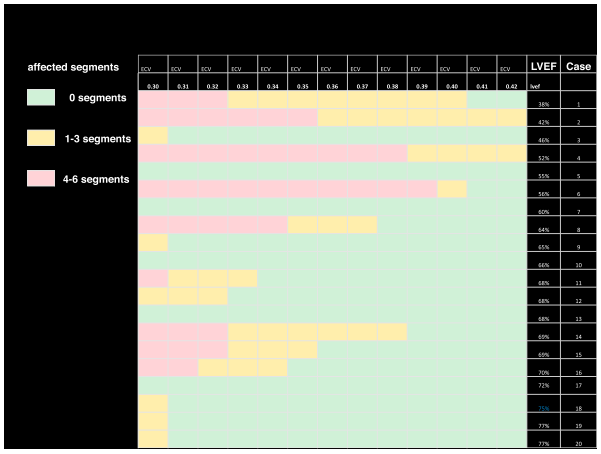
Table.





P2856 - TETRALOGY OF FALLOT – LESSONS LEARNED FROM PRENATAL DIAGNOSIS. 15 YEAR EXPERIENCE OF A TERTIARY REFERENCE CENTER

Agnieszka Grzyb[1], *Adam Koleśnik*[1], *Renata Bokiniec*[2], *Marzena Dębska*[3], *Joanna Szymkiewicz-Dangel*[4]
Warsaw Hospice for Children Foundation, Reference Center for Fetal Cardiology, Warsaw-Poland[1]; *Medical University of Warsaw, Neonatology And Neonatal Intensive Care Department, Warsaw-Poland*[2]; *The Center of Medical Postgraduate Education, Department of Obstetrics And Gynecology, Warsaw-Poland*[3]; *Medical University of Warsaw, Reference Center For Fetal Cardiology, 2nd Department of Obstetrics And Gynecology, Warsaw-Poland*[4]

Background: Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease. Detailed information from prenatal diagnosis help to ensure appropriate perinatal management.
Materials and Methods: We evaluated echocardiographic examinations and perinatal follow-up data of fetuses with TOF diagnosed between 2002–2016 in the reference center for fetal cardiology.
Results: TOF was diagnosed in 239 fetuses (M/F≈1): 169 with pulmonary stenosis, 58- pulmonary atresia (TOF-PA; 5 evolving from stenosis) and 12- absent pulmonary valve (TOF-APV). The number of cases increased yearly (3-38 cases), with simultaneous drop in the median time of diagnosis (32-22 weeks). Karyotype was checked in 180 fetuses and showed abnormalities in 39 cases, mostly trisomy (T21-n = 20, T18-n = 4, T13-n = 7). Molecular examinations (FISH or microarray) were performed in 128 fetuses and found anomalies in 28, including 26 cases of microdeletion 22q11 (most common in TOF-APV). Hypoplastic thymus, right aortic arch (RAA) and polihydramnios were indicative of DiGeorge syndrome and AVSD- of T21 ($p < 0.05$). Coexisting anomalies were common, both cardiac (45%, most often: RAA, AVSD, LSVC) and extracardiac (47%, most often: hypoplastic thymus, urinary and skeletal anomalies; small for gestational age-6%). Ductus arteriosus was absent in all TOF-APV fetuses and in single cases of TOF and TOF-PA, significantly affecting perinatal management. Follow-up was known in 211 cases, most children were live-born (n = 176, 83.4%) in good condition, except for TOF-APV (only 1 child living). Stillbirths (n = 20, 9.5%) and terminations of pregnancy were rare (n = 15, 7%), all in cases with coexisting serious genetic or structural anomaly or severely hypoplastic pulmonary arteries.
Conclusions: Introducing 3-vessels-trachea-view into screening ultrasound improved prenatal detection of TOF. Fetuses with TOF require detailed anatomy-focused ultrasound scan and genetic examination (possibly microarray). Thymus hypoplasia is a specific marker of microdeletion 22q11. Repeated examinations are important to assess progression of RVOTO. Perinatal outcome is usually good (except for TOF-APV fetuses), however significantly affected by coexisting anomalies.

P2859 - FETAL DIAGNOSIS OF COMPLETE VASCULAR RING IN RIGHT AORTIC ARCH AND LEFT DUCTUS ARTERIOSUS

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Ospedale Pediatrico Bambino Gesù, Perinatal Cardiology, Rome-Italy[1]; *Ospedale Pediatrico Bambino Gesù, Cardiology, Roma-Italy*[2]; *Ospedale Pediatrico Bambino Gesù, Cardiac Surgery, Roma-Italy*[3]; *Ospedale Pediatrico Bambino Gesù, Radialogy, Roma-Italy*[4]

Background: Right Aortic Arch with Aberrant Left Subclavian Artery and Left Ductus Arteriosus (RAA + ALSA + LDA) is a common type of vascular rings. Clinical presentation is subtle and post-natal echocardiographic diagnosis is often challenging. Fetal ultrasound, with visualization of wide open LDA, can make identification of this anomaly easier. We reviewed our experience of pre and postnatal diagnosis of RAA + ALSA + LDA.
Methods: Data from all patients with diagnosis of RAA + ALSA + LDA between 2010 and 2014 referred at our tertiary centre were reviewed. All patients underwent CT and/or MRI for airway compression. Indication for surgery was the presence of symptoms and/or tracheal compression (as defined as a narrowing of the tracheal lumen ≥ 50% than the maximum proximal diameter) and/or tracheomalacia.
Results: we observed 52 consecutive patients with RAA + ALSA + LDA. Prenatal diagnosis was made in 26 (50%) patients. Five were lost at follow-up. Six (11.5%) patients had associated Ventricular Septal Defect (VSD), 1 (2%) atrioventricular septal defect, 1 (2%) Ebstein anomaly and 1 (2%) Tetralogy of Fallot (TOF). De George Syndrome was present in 6 (11.5%), of them 2 (4%) had VSD and1 (2%) TOF. One patient (2%) had CHARGE and 1 (2%) Goldenhar Syndrome. Relevant tracheal compression and/or tracheomalacia was identified in 14/42 patients (33%), of which 50% were symptomatic. Overall 15/42 (36%) had clinically evident symptoms. Surgery was performed in 22/42 (52%) patients. One of them had persistent tirage. Patients with prenatal diagnosis had lower incidence of symptoms and tracheal

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compression with/out tracheomalacia than those with postnatal diagnosis (22% vs 44%, 44% vs 67%, p = NS).
Conclusion: RAA + ALSA + LDA has a good prognosis and mostly asymptomatic. Compression and/or malacia are not always associated with symptoms, potentially increasing the number of missed diagnosis. Since post-natal echocardiographic diagnosis is challenging, prenatal identification of this anomaly is essential for the early management and to avoid irreversible damage of the upper airways.

P2861 - CAN SIMPLE ECHOCARDIOGRAPHIC RIGHT VENTRICLE DIAMETERS Z SCORE PREDICT RIGHT VENTRICULAR ENLARGEMENT IN CONGENITAL HEART DISEASE

Luisa Garcia-Cuenllas Álvarez[1], Ana Rodriguez-López Domingo[2], Federico Gutierrez Larraya[3], Enrique Maroto Álvaro[4], Constancio Medrano López[4]
Hospital Universitario De Salamanca, Pediatric Cardiology, Salamanca-Spain[1]; Hospital Hospiten, Cardiology, Tenerife-Spain[2]; Gregorio Marañón, Pediatric Cardiology, Madrid-Spain[3]; Hospital Universitario Gregorio Marañón, Pediatric Cardiology, Madrid-Spain[4]

Introduction: echocardiographic measurement of RV diameters represents an easy, fast and reproducible way to quantify this chamber. Reference values and Zscores of RV diameters are provided. Accurate assessment of RV size is important in treatment and follow up of congenital heart disease (CHD). Cardiac magnetic resonance (CMR) is the gold standard to measure RV dilatation. Disagreement between linear measurements and CMR volumes (indexed RV end-diastolic volume, iRVEDV) had been proved, but not between Zscore and CMR.
Methods: 700 healthy children (0 days to 18 years old) were enrolled. RV end-diastolic diameters (basal, midcavity and longitudinal) were measured and indexed using body surface area (Haycock formula). Zscore were computed according to literature requirements. These parameters were validated against 50 post-operated Tetralogy of Fallot patients (PTF) iRVEDV obtained by CMR.
Results: Exponential and square root regression models resulted in the best fit with R2 of 0.81, 0.82 and 0.9 respectively for each diameter. They all satisfied the assumption of homoscedasticidy and normality of residuals (Bresch-Pagan and Saphiro-Wilk tests). Confounders as gender and inter/intraobserver variability were considered. Predicted Z scores of basal, midcavity and longitudinal diameters plus plots of standardized residuals are presented: Image 1

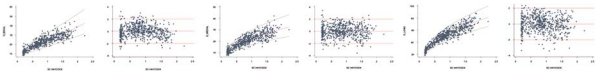
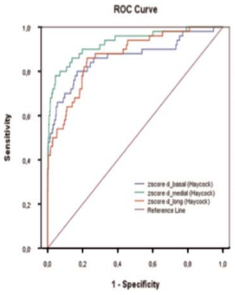


Image 1.

There was a weak correlation between diameters and RVEDVi (Rho Spearman < 33%). But diameter´s Zscore in ROC curves showed a very good discriminating capacity with AUC >0.8, able to determine which RV were normal sized and which were not (dilated PTF, iRVEDV >110 ml/m2). A cut-off point of > + 2 Zscore provided high specificity (>97%). Image 2
Conclusions: reference values for RV diameters represent a valid diagnostic tool for RV quantification in children. RV diameter´s Zscores could discriminate between normal and dilated RV. Diameter´s Zscores could predict RV enlargement in PTF patients. We offer an easy tool to guide follow up and decision making relating to RV dimensions in healthy and CHD children.



Z-score	AUC (area under curve)	IC 95%	
Basal diameter	0.86	0.79	0.93
Medial diameter	0.93	0.88	0.97
Longitudinal diameter	0.87	0.82	0.92

Image 2.

P2867 - THE AORTA IN TURNER SYNDROME IT'S NOT ALL ABOUT THE SIZE

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Background: Aortic dissection occurs more frequently in TS than in the general population, at younger ages and smaller aortic sizes, suggesting that aortic dilation is not the only etiology. We hypothesized that aortic stiffness indices in TS, as measured by echo, differ from normal controls (NC).
Methods: 102 TS subjects and 88 age matched healthy NC females were prospectively enrolled. 2-dimensional and M-mode echo images of the ascending aorta were acquired using a standard protocol. Aortic stiffness (ASI), aortic distensibility index (ADI) and elastic modulus (EM) for the ascending aorta were calculated. TS subjects were grouped as "with risk" if one or more of the following risk factors were present: bicuspid aortic valve, coarctation, hypertension, hyperlipidemia or diabetes mellitus (n = 60), and as "no risk" (n = 42) if none were present. The 3 groups were

Table.

Variable	GROUP NC n = 88	TS n = 102	P-Value
Age (years)	37.9 ± 13.8	34.3 ± 16.3	0.100
Body Surface Area (m^2)	1.7 ± 0.2	1.6 ± 0.3	**<0.001**
Body Mass Index (kg/m^2)	25.6 ± 14.7	27.9 ± 7.1	0.171
Heat Rate (bpm)	65.9 ± 10.2	81.1 ± 15.5	**<0.001**
Systolic Blood Pressure (mmHg)	119.5 ± 13.2	120.3 ± 19.3	0.755
Diastolic Blood Pressure (mmHg)	70.9 ± 9.4	69.7 ± 10.2	0.400
Aortic root- 2D (cm)	2.6 ± 0.3	2.7 ± 0.3	**0.034**
Ascending Aorta- 2D (cm)	2.8 ± 0.3	2.7 ± 0.4	0.764
Ascending Aorta-M mode (cm)	2.8 ± 0.4	2.7 ± 0.5	0.064
ADI -M-mode ($mmHg^1.10^{-3}$)	7.1 ± 3.6	5.3 ± 2.7	**<0.001**
2 (Aorta systole - Aorta diastole)/ Aorta diastolex(systolic blood pressure-diastolic blood pressure)			
ASI-M-mode (no unit)	3.9 ± 2.2	5.3 ± 2.9	**<0.001**
[In (systolic blood pressure/diastolic blood pressure)]/[Aorta systole-Aorta diastole/Aorta diastole].			
Ascending aorta EM -M mode (kilo -pascal)	49.7 ± 26.8	66.2 ± 39.6	**0.002**
[(Systolic blood pressure- diastolic blood pressure)x Aorta diastole] / (Aorta systole-Aorta diastole)			

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compared using ANOVA; individual groups were compared using Student's t –test. A multivariate regression model estimated independent group associations after adjusting for age and aortic size. Pearson correlation coefficients were used to assess the correlation of stiffness indices with systolic blood pressure.
Results: Table shows the comparison between TS and NC. TS subjects had larger aortic root and aortic size indexed to BSA ($p < 0.001$) but comparable sized ascending aorta as NC. Ascending aorta ASI, ADI and EM (M-mode) differed significantly between TS and NC ($p < 0.001$); this difference persisted when adjusted for age and aortic size. Aortic stiffness indices did not differ between the two TS risk groups ($p > 0.17$). ASI and EM correlated positively with SBP ($r = 0.3$ and $r = 0.49$ respectively) while ADI correlated negatively ($r = -0.41$).
Conclusion: Women with TS have stiffer and less distensible ascending aortas when compared to age matched NC. This difference is independent of aortic dilation and previously described risk factors, suggesting that the aorta may be innately abnormal in TS.

P2868 - FETAL DIAGNOSIS OF SUBAORTIC OUTFLOW TRACT OBSTRUCTION WITH AORTIC ARCH COARTATION OR INTERRRUPTION DIAGNOSIS AND MIDTERM POSTNATAL OUTCOME

Queralt Ferrer[1], Alberto Rafael Guerra[2], Silvia Arevalo[3], Gemma Giralt[2], Anna Sabate-Rotés[2], Ferran Gran[2], Irene Ribera[3], Joaquin Fernandez-Doblas[4], Raul Abella[4], Ferran Roses[2]
Vall Hebron, Pediatric Fetal Cardiology, Barcelona-Spain[1]; Vall Hebron, Pediatric Cardiology, Barcelona-Spain[2]; Vall Hebron, Fetal Medecine, Barcelona-Spain[3]; Vall Hebron, Pediatric Cardiac Surgery, Barcelona-Spain[4]

Introduction: Borderline hypoplastic left heart syndrome is an inaccurate term that includes a whole spectrum of left heart hypoplasia. Due to the unique nature of the foetal circulation, prenatal diagnosis of subaortic left ventricular outflow tract obstruction (SLVOTO) in context of borderline left heart structures is very difficult and there are no specific definitions. We present clinical outcome of foetuses with SLVOTO with either Aortic arch hypoplasia + Coarctation or Interrupted aortic arch (IAA).
Methods: We retrospectively reviewed 19 foetuses with SLVOTO, defined as subaortic left ventricular outflow tract <4mm(measured at 32 GW), in fetuses with either Aortic arch hypoplasia + Coarctation or IAA between Jan-2010 and Oct-2016. Main Foetal echocardiography mesures were: left ventricle diastolic diameter, mitral annulus, aortic annulus and ascending aorta and Isthmus z-scores and subaortic LVOT in mm.
Results: Out of 19 foetuses, 12 were females(63,2%), with a medium gestational age at time of CHD diagnosis of 26 GW. 16 (84%) had Aortic arch hypoplasia + Coarctation and 3(16%) IAA. Of all 19 foetuses, 9(47%) had a large unrestrictive VSD. 14(74%) foetuses had mitral valve annulus z-score < -3. All had neonatal surgical biventricular repair without hospital mortality. Interestingly, out of 5 foetuses with severe SLVOT < 3 mm, 4(80%) required subaortic stenosis surgical relieve, whereas out of 14 with SLVOT >3mm, only 1(7,2%) required postnatal subaortic surgical relieve ($p = 0.02$). After a medium follop-up of 2.2years (3months-7years), all 19 patients were alive. 2 patients developed late subaortic stenosis, 26% developed mild-moderate mitral stenosis. 10% had mild aortic valve stenosis and 26% required aortic arch balloon dilation . In addition, 2 patients had post-operative complete heart block that required pacemaker implantation.
Conclusions: Severe Prenatal diagnosis of SLVOTO in patients with either Aortic arch hypoplasia + Coarctation or IAA is a risk factor for postnatal subaortic surgical relieve but still all foetuses underwent a biventricular repair with no mortality.

P2876 - FETAL ATRIAL SEPTAL ANEURYSM POSTNATAL OUTCOME IN A SERIES OF 29 CASES

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Background: Atrial Septal Aneurysm (ASA) has been associated with cardiac defects including atrial septal defects, atrioventricular valve obstruction, pulmonary venous obstruction and arrhythmias. It has also been identified prenatally as an isolated finding, in which the significance is not clearly known.
Materials,Methods: Retrospective study including 29 fetuses with ASA diagnosed by fetal echocardiography between 2011 and 2016. Fetal ASA was defined as a redundant atrial septal tissue extending at least halfway across the left atrium diameter. Data on prenatal, perinatal and postnatal outcome was reviewed.
Results: ASA was diagnosed in 29 fetuses at a mean gestational age (GA) of 33.2 weeks (range 21.3–37.3weeks). The main indication for fetal echocardiography was the suspicion of an abnormal foramen ovale on screening ultrasound. Associated extracardiac anomalies were detected in one case. However, nineteen fetuses (66%) presented other cardiac anomalies:1 fetal arrhythmia, 1 mild pulmonary stenosis, 1 perimembranous ventricular septal defect (VSD), and 15 cases with significant right heart dominance (defined by a ratio of right-to-left cavities greater than 1.4). Overall there were 10 fetuses with isolated fetal ASA. A chromosomal anomaly (46,XY,del(13)(q13q14)) was identified in one fetus with associated perimembranous VSD, and parents opted for termination. The remaining cases were confirmed to be chromosomally normal either pre or postnatally. Perinatal outcome was normal in all newborns.
Postnatal outcome: Postnatal follow-up was available for 28 newborns. Mean follow-up was 29 months (range 4-60 months). Pulmonary hypertension and mitral stenosis was diagnosed in one case with right heart dominance. Aortic coarctation was not confirmed in any case. Atrial-septal defect was diagnosed in 11 cases after delivery (39%), of which 7 closed spontaneously before one year of life.
Conclusions: In our series, fetal ASA was not associated with pulmonary venous obstruction, significant neonatal arrhythmias or aortic coarctation. However, our results support the need for postnatal echocardiography in order to exclude atrial-septal defects.

P2879 - ECHOCARDIOGRAPHIC PREDICTORS OF COIL VERSUS DEVICE CLOSURE IN CHILDREN UNDERGOING PERCUTANEOUS PDA CLOSURE

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Introduction: The need to determine whether a PDA could be closed by coil or device remains an important issue in planning the mode of closure especially in developing countries where the

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availability and cost of such devices are important factors in decision making.
Patients and Methods: This study included 66 patients who were referred for elective trans-catheter closure of PDA. All patients underwent full echocardiogram before closure to measure the pulmonary arterial end diameter, PDA color flow width and extent, the peak and diastolic Doppler gradients across the duct, diastolic flow reversal across the descending thoracic aorta, Qp/Qs, left atrial dimensions and volume, left ventricular sphericity index, left ventricular dimensions, volumes and systolic function. The patients were then subdivided into 2 groups based on the mode of closure whether by coil (n = 37) or device (n = 29) and the different parameters measured were compared between the 2 groups.
Results: Using univariate analysis the device closed group had statically significant higher end systolic and diastolic volumes indexed measured by both M-mode and modified Simpsons method, Left atrial volume, Qp/Qs, pulmonary end diameter measured from both the ductal view and parasternal short axis view, color flow width, color flow extent and diastolic flow reversal. Using multivariate analysis the independent predictors for Device closure included: PDA pulmonary end >2.5 mm measured from supra sternal view, Color flow width of PDA jet >2.3, Diastolic flow reversal time >96.6% of the entire diastole, color flow extent of PDA jet in the main pulmonary artery >70% of pulmonary artery length and Qp/Qs >1.5.
Conclusion: The study concluded that the selection between coil or device closure methods can be done on the basis of non-invasive anatomic and hemodynamic data obtained by 2D and Doppler echocardiography.

P2884 - THE DISTRIBUTION OF CONGENITAL HEART DISEASES IN RELATIONSHIP WITH ALTITUDE

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Introduction: Congenital heart diseases (CHD) have a direct incidence increase with altitude. Aim of the study: To analyze the occurrence of the most frequent CHD at different altitudes.
Method: A team of pediatric cardiologists performed echocardiographic studies in children with suspicion of CHD in different cities of Bolivia during missions, in places where there is no pediatric cardiology service. The patients were distributed in two groups: One: Living at more than 3600 m and two: Living below 3000 m. The protocol was unique and the examinations were conducted by a single echocardiographist.
Results: Between 2008 and 2016 4453 patients were examined; Group 1: 3179 patients and Group 2: 1274. 340 Patients with trisomy 21 were included and represented 10,7% of the total number. In group 1 complete AV channel represented only 2,4% of the total number. At high altitude 11,6% Patients showed pulmonary hypertension in absence of CHD, this number decreases at lower altitude. In Group 1: We examined 3179 patients. The VSD together with the ductus are the most frequent diseases. The isolated pulmonary arterial hypertension was present in % patients. In Group 2 the VSD is the most frequent disease followed by ASD. Speaking of severe CHD In group 1 tricuspid atresia and Ebstein malformation are the most common severe CHD, transposition of great arteries and hypoplastic left heart showed a very low percentage in the occurrence. This number increase in inverse proportion with altitude. In group 2 we founded hypoplastic left heart syndrome as the most seen severe CHD.
Conclusions: Hypoxic Hypobaric Hypoxia has an influence in the distribution of CHD and in the occurrence of pulmonary hypertension.

P2890 - MORPHOLOGY OF PULMONARY ARTERIES IN PULMONARY ATRESIA WITH INTERVENTRICULAR SEPTAL DEFECT A VISION OF COMPUTED TOMOGRAPHY ANGIOGRAPHY

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Introduction: Pulmonary atresia with ventricular septal defect (PA + VSD) is a rare cardiac malformation, comprising a complex variation of pulmonary artery morphology and pulmonary arterial supply. Correct identification of this anatomy is essential for treatment planning.
Objective: Study the different subtypes of pulmonary arterial supply through computerized angiography in children with PA + VSD applying the classifications described by Jane Somerville (1970) and Barbero-Marcial (1990) and their applicability in treatment orientation.
Material and Methods: We reviewed the medical records of 47 children with PA + VSD, of a children's cardiology service at a tertiary center. The volumetric data acquisition was performed after venous contrast, in a Siemens 64 detector row scanners with 80 / 100kv, 60/100 mAs and thickness of 0.6 mm.
Results: 55.5% of the patients were male (26 patients), with a mean age of 5.1 ± 5.24 years. According to Jane Somerville's classification, 11% of the patients belonged to subgroup I, 52% to subgroup II, 26% to subgroup III and 11% to subgroup IV. In the Barbero-Martial classification, 26% of the cases are of group A, 63% of group B and 11% of group C.
Conclusions: CT angiography is a noninvasive method able to identify all morphological arteries subtypes within the classifications proposed by Jane Somerville and Barbero-Marcial. The results showed concordance between the classifications being the most common subtype B, in 63% of the cases (B3 the most frequent) according to the Barbero-Marcial classification and the subgroups II/III in 78%, according to Jane Sommerville. These findings facilitate the surgical decision in the approach of collateral, reducing the need for invasive angiography.

P2896 - THE IMPACT OF CORONARY ANGIOTOMOGRAPHY IN THE EVALUATION OF KAWASAKI DISEASE

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Introduction: Kawasaki disease (KD) is an acute, self-limiting vasculitis that affects infants and young children. Giant coronary aneurysms are the most serious complication, with a mortality rate reaching 25% of untreated patients and 4% of treated ones. Even patients with giant aneurysms may be asymptomatic. The echocardiogram is the routine examination on the follow-up of these patients, since the initial portion of the coronary arteries can be analyzed in younger children. However, coronary angiography (CT angiography) is a more accurate method of evaluating all the coronary branches, diagnosing dilatation, stenosis and calcification even of the most distal vessels.

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Methods: Retrospective analysis of patients with a diagnosis of KD in outpatient follow-up at Instituto Nacional de Cardiologia. Descriptive analysis of the findings of echocardiography, CT angiography and functional class.
Results: Five patients were evaluated, 4 male and 1 female, aged 4 to 13 years. Four received immunoglobulin and platelet aggregation inhibitor during the acute fase. The echocardiogram showed dilatation or aneurysm of the proximal left coronary artery in all patients and some involvement of the right coronary (RC) artery in 3 cases. All patients underwent CT angiography in a 64-channel CT scanner with cardiac trigger and venous beta-blocker. Thrombus and occlusion of the anterior descending (AD) artery was seen in 3 patients. In the circumflex artery we observed thrombus and occlusion in one case and dilation without occlusion in another case. One patient had RC occlusion and another had RC stenosis. All are in clinical follow-up and NYHA functional class 1.
Conclusion: Studying our series, we acknowledged that CT was better than echocardiography due to the possibility of three-dimensional evaluation, precise quantification of the lesion even in distal branches and identification of intraluminal thrombi. Although exposure to ionising radiation and contrast are still a concern with the method, it was decisive to the clinical decision.

P2898 - CARDIAC RHYTHM DISTURBANCES PRENATAL DIAGNOSIS AND FETAL POSTNATAL OUTCOMES

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Objectives: The aims were to investigate the prevalence of fetal arrhythmias, the maternal/ fetal risk factors involved and the postnatal outcomes.
Methods: From 06/2011 to 03/2016, 1984 pregnant women were prospectively investigated, considering the following groups of variables: diagnostic investigation, hemodynamic/functional repercussions, risk factors and evolution.
Results: Rhythm disturbances in 267 fetuses showed different distribution along the gestational age (GA): atrial flutter (AF) and supraventricular tachycardia(SVT) in 6.0% and 11.0%, respectively (26-28w); congenital atrioventricular block (CAVB) in 9.0%, before 24w; transient sinus taqui/bradicardia (TST/B) with regular distribution, and premature atrio/ventricular contraction (PAVC), were seen in 71,2%, before 24w and after 33w and tri/bigeminy (26-28w) in 3.0%. TST was related with fetal anemia, fetal/maternal infection and maternal intake of thermogenic foods ($p < 0{,}005$); SVT was associated with Ebstein anomaly and complete CAVB with isomerism and maternal autoimmune antibodies ($p < 0{,}005$). Isolated pericardial effusion was present in 74/267 and hydrops in 22/267; 8/36 with TST showed cardiomegaly/pericardial effusion and 1/36 hydrops, all of them with anemia. More severe repercussions were seen in those with AF or complete CAVB (pericardial effusion:88.0%; hydrops and/or cardiomegaly: 60.0%) and SVT (pericardial effusion:86.7%; hydrops and/or cardiomegaly:49.7%). Among patients with SVT or AF, the majority improved with monoterapy (29/52) and 17/52 demanded multitherapy; there were no deaths and eight were preterm due to fetal arrhythmia with hydrops. Among those with complete CAVB and complex congenital heart disease ($n = 8$) there were seven intrauterine deaths and one postnatal pacing; no death was seen in the group of immune-mediated damage ($n = 16$): 12.5% had premature deliveries and 43.8,% postnatal pacing. One patient of unknown aetiology was born at term without complications.
Conclusions: Most arrhythmias are benign, but SVT, AF and CAVB were associated with poor prognosis and high mobimortality. The continuous prenatal surveillance is important to make

P2901 - ECHOCARDIOGRAPHIC QUANTIFICATION OF SYSTOLIC FUNCTION IN THE SINGLE SYSTEMIC RIGHT VENTRICLE CORRELATION WITH MAGNETIC RESONANCE IMAGING DERIVED EJECTION FRACTION

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Background: Preservation of systolic function is of paramount importance in patients with single systemic right ventricles (RV). Ejection fraction (EF) is considered the most useful measurement of systolic function but is difficult to calculate by transthoracic echocardiography (TTE). Alternative TTE parameters have been proposed, however contraction patterns of the systemic RV differ from the biventricular heart. Our objective was to determine which TTE parameters best reflect systemic RV EF.
Methods: Subjects with hypoplastic left heart syndrome (after bidirectional or total cavopulmonary anastomosis) were prospectively recruited. Cardiac magnetic resonance imaging (cMRI) was performed and RV EF calculated. TTE was performed at a median interval of 1 day from cMRI; parameters of systolic performance were assessed as outlined in Results. Biplane fractional area change (FAC) was obtained by averaging apical and short axis FAC; triplane FAC included the RV inflow/outflow view. Spearman's rank correlation was used for comparisons between cMRI-derived EF and TTE measurements.
Results: A total of 59 cMRI/TTE pairs from 45 subjects were analyzed. Median age was 10.2yrs (range, 9mos -27.9yrs). Results with absolute rank correlation values >0.6 are summarized in the Table; correlation was <0.6 for global longitudinal systolic strain, TAPSE, MPI, IVA, and peak tricuspid lateral annulus systolic velocity.
Conclusion: Many TTE parameters of RV function did not correspond well to cMRI-derived EF in the single systemic RV. Quantitative parameters with the most promising correlations included FAC, particularly biplane FAC, as well as short axis values for circumferential strain and RV-end systolic area. In the systemic RV, short axis views may provide more robust quantitative TTE assessments of systolic function, given the altered geometry and contraction patterns of the single systemic RV. No one TTE parameter correlates optimally with EF, therefore research into a multivariable algorithm may be beneficial.

Table.

	Mean ± SD	P-value	Spearman's Rank coefficient
CMR			
Ejection Fraction	45.2 ± 9.1%	-	-
TTE Variables with Correlation > 0.6			
Biplane FAC	41.1 ± 8.4%	<0.001	0.69
Triplane FAC	38.7 ± 6.7%	<0.001	0.68
Short axis circumferential strain (VVI)	14.3 ± 5.2%	<0.001	−0.68
Short axis FAC	43.4 ± 10.7%	<0.001	0.66
Short axis indexed RV end-systolic area	11.8 ± 6.1 cm^2	<0.001	−0.64
Visually estimated EF	50.0 ± 9.3%	<0.001	0.63

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P2902 - THE IMPORTANCE OF EARLY DIAGNOSIS OF THE ANOMALOUS ORIGIN OF THE CORONARY ARTERY

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Introduction: The anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare disorder, estimated at 1:300,000 live births, comprising 0.24% to 0.46% of all congenital heart defects. These number might be underestimated because the diagnosis is very difficult. Early diagnosis and treatment may be crucial for the prognosis of these patients.
Objectives: We aim to report a series of 10 patients with various clinical presentations related to the diagnosis of ALCAPA undergoing treatment between April 2011 and 2016, at the same institution, describe their treatments and results and analyze their clinical evolution.
Methods: Ten patients, 7 females and 3 males, aged 16 days to 14 years at the time of surgery and weighing between 3.2 and 50 kg were analyzed retrospectively and evaluated according to their signs and symptoms in the time of diagnosis and during their clinical evolution.
Results: Of the patients evaluated, 3 presented only systolic murmur, 5 had also dyspnea and symptoms of heart failure, 1 had unstable angina and 1 cardiogenic shock and need of mechanical ventilation. The 10 patients assessed had ALCAPA. Nine patients underwent surgical treatment, with 7 cases of coronary reimplantation, one exclusion of the anomalous vessel, one case of coronary artery bypass from the anterior descending artery (DA) with DA ligation in the pulmonary trunk and 1 patient died before surgery. Follow-up ranged from 6 months to 5 years. Seven patients remained asymptomatic, 5 of which with supravalvar pulmonary stenosis, 1 with pulmonary insufficiency and 1 with no residual lesion. Two patients died (1 in the preoperative and 1 in the immediate post operative period).
Conclusion: Of the 9 patients submitted to surgery, 8 had complete remission of symptoms. Early diagnosis is vital, so it's important to remember ALCAPA when evaluating heart murmurs, myocardial ischemia and myocardial dilation of unknown ethiology.

P2904 - PRENATAL DIAGNOSIS OF COARCTATION OF AORTA ECHOCARDIOGRAPHIC PREDICTORS

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Introduction/ Objectives: The prenatal diagnosis of CoAo is challenging and can be suspected when there is cardiac ventricular or great vessels disproportion. The sensitivity and specificity of several criteria have a high false positive rate. The aim was to establish the echocardiographic (ECHO) diagnose based on predictors for the prenatal diagnosis of coarctation of aorta (CoAo).
Methods: The study included pregnant women referred for fetal ECHO between June 2011 and December 2015 due to ventricular and/or great vessels asymmetry. The fetuses were scanned from 28 to 38 weeks (mean = 32 ± 4 weeks). Fetal ECHO parameters for suggestive cases were: right/left ventricular (RV /LV) diameters ratio ≥1.7, main pulmonary artery/ascending aorta (MPA/AAo) ratio ≥1.7, z-score of ascending aorta (AAo) ≤ − 1.5 and z-score of aortic isthmus ≤ − 1.5. Multivariable analysis and logistic regression were used to assess the interdependence of significant variables.
Results: A total of 52 women were scanned and 28 had criteria for prenatal suspicion of CoAo. The CoAo was confirmed after birth in 19 fetuses (67.9%). The isthmal Z-score ≤ − 1.5 and MPA/AAo ratio ≥1.7 demonstrated a high sensitivity (88% and 82%) and positive predictive value (79 and 77%). Although ventricular asymmetry and AAo Z-score had great sensitivity (74% and 76%) they had poor specificity (34% and 32%). The association of two or more parameters showed 94% of accuracy. Nine patients with mild/moderate hypoplastic aortic arch did not develop CoAo during the follow up of one year. Those were considered false-positive diagnoses.
Conclusions: Antenatal diagnosis of CoAo is critically important for improved preoperative hemodynamic stability of the neonate and reduced mortality. Prenatal diagnosis of aortic coarctation shows high false-negative rates at screening and poor specificity by fetal ECHO . A combination of echocardiographic parameters can improve the likelihood of a correct diagnostic.

P2908 - CONVENTIONAL ECHOCARDIOGRAPHIC PROFILE AND BY MEANS OF STRAIN IN PATIENTS WITH MUCOPOLYSACCHARIDOSIS IN BAHIA BRAZIL

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Background: Mucopolysaccharidosis (MPS) is a genetic disease, a lisossomal storage of glycosaminoglycans. It affects various organs, with frequent cardiovascular compromise, characterized mainly by left valvular lesions and left ventricle (LV) hypertrophy. The cardiovascular signs and symptoms are underestimated due to the simultaneous involvement of the disease in other organs. Enzyme replacement therapy (ERT) can be used in MPS I, II, IV and VI. In face of this scenario, the knowledge of the application of new tools of conventional and advanced echocardiography is relevant in order to improve cardiac care.
Materials and Methods: This is an outpatient sectional descriptive study, from the genetic services of the Federal University of Bahia-Brazil. The patients underwent conventional echocardiography and strain by speckle-tracking from January to June, 2016.
Results: 16 patients have been evaluated with median age of 14.2 years (deviation: 5.2 years); 12 (75%) were male. The most common type: MPS VI (8 patients - 50%), MPS II (4 patients - 25%), MPS I (3 patients - 18.8%) and MPS III (1 patient - 6.2%). Left valve lesion was found in 15 patients (93.3%) with a higher prevalence of mitral lesions - 13 patients (81.2%) all of which had insufficiency. Twelve (75%) showed concentric LV remodeling. All patients had LV systolic function preserved (Simpson and Teichholz). Three (18.8%) patients had abnormal LV MPI (myocardial performance index). Nine (56.2%) patients had change in LV global longitudinal strain (SGL). The study showed a positive association between mass and change the LV MPI and LV SGL and start time of ERT and LV MPI and LV SGL.
Conclusions: Echocardiographic changes in patients with MPS are frequent, especially the left valve changes, change in LV geometry and subclinical LV dysfunction. The use of the new tools of conventional and advanced echocardiography can improve this follow-up.

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P2914 - FIBROUS SKELETON LESION DIAGNOSTIC AND THERAPEUTIC CHALLENGE

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Introduction: The heart's fibrous skeleton consists of dense connective tissue that surround the base of the semilunar and atrioventricular valves. The connective tissue that separates the atria and ventricles comprises the mitral, tricuspid and aortic valves designing a fibrous triangle.
Case Report: Boy diagnosed at birth with perimembranous ventricle septal defect (VSD) and subaortic membrane. Cardiac catheterization was performed at one year of age showing a left ventricular outflow gradient of 50 mmHg. At 2 years of age, surgery was performed to close the VSD and resect the membrane. He was again referred to surgery after 9 years due to severe aortic regurgitation. A repair the right coronary leaflet was unsuccessful, therefore he underwent a metallic aortic valve replacement surgery. Post operative showed moderate aortic regurgitation. After 3 years, he presented dyspnea and chest pain. With progressive worsening of symptoms, the aortic valve was replaced after 1 year. Postoperative echocardiogram showed a mild regurgitation and a maximum antegrade gradient of 30 mmHg. After another year, he presented with fever, weight loss and dyspnea. Infective endocarditis was ruled out. Echocardiography showed pulmonary artery hypertension, right ventricular overload and a rupture of the anterior leaflet of the mitral valve. He then underwent a successful surgery and has been clinically well.
Discussion: Patients with previous manipulation of the fibrous triangle should be cautiously examined due to risk of lesions requiring complex techniques of diagnosis and therapy.

P2917 - EVALUATION OF AORTIC ANOMALIES IN THE NEONATAL PERIOD THROUGH ANGIOTOMOGRAPHY

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Introduction: The hemodynamic severity and clinical manifestations of aortic malformations range from mild narrowing or tortuosity of the aortic isthmus in an asymptomatic patient to severe obstruction associated with cardiogenic shock. In addition to clinical symptoms, several imaging methods have been used to assess the severity of these malformations. Aortic angiotomography is able to evaluate the entire aortic arch with high spatial resolution, and in the newborn it is essential to complement the echocardiogram in cases in which the acoustic window does not allow evaluation of the entire anatomy.
Methods: We report the cases of 7 neonates (3 males and 4 females) with malformations of the aorta, aged between 8 and 27 days. At admission, all of them had clinical signs of congestive heart failure and were on intravenous prostaglandin. The examinations were performed on a 64-channel SIEMENS scanner under general anesthesia. The aortic angiotomography imaging aided the therapeutic approach, adding information to the echocardiogram. Interruption of the aortic arch was observed in 3 cases, coarctation of the complex aorta in 3 cases and aortic hypoplasia in one case. Associated malformations were: atrial septal defect, ventricular septal defect, right ventricular outflow tract obstruction, patent foramen ovale, patent ductus arteriosus, single ventricle and mitral agenesis. All patients underwent surgical correction after angiotomography.
Results: Diagnosis by transthoracic echocardiography alone may present some limitations. The chest angiotomography was able to evaluate the ascending, transverse and descending aorta, vessels of the base with their variations, besides evaluating and quantifying the presence of collateral circulation. Nowadays, with low-dose radiation protocols, diagnostic complementation with angiotomography should be part of the preoperative approach.

P2923 - IMPACT OF VENTRICULOTOMY ON THE SYSTEMIC RIGHT VENTRICLE IN HYPOPLASTIC LEFT HEART SYNDROME

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Background: In hypoplastic left heart syndrome (HLHS), right ventricle (RV) to pulmonary artery shunts (RVPAS) have been increasingly used during stage I palliation. RVPAS requires an RV incision, whereas modified Blalock-Taussig shunt (MBTS) does not, leading to concerns about long term effects of ventriculotomy on the systemic RV after RVPAS. This study evaluated for RV myocardial changes after RVPAS by cardiac magnetic resonance imaging (cMRI) and transthoracic echocardiography (TTE).
Methods: Subjects with HLHS were prospectively recruited and underwent cMRI and TTE. A subset of patients with prior RVPAS was compared to an age-matched group with prior MBTS.
Results: Twenty subjects with prior RVPAS participated. Median age was 2.0 years (range, 9 months - 11 years). Thirteen were post-Stage II palliation and 7 were post-Stage III. By cMRI, all 20 subjects had a thinned, dyskinetic section of the anterior RV wall

Table 1. Comparison of Age-and Stage-Matched HLHS Patients with History of MBTS of RVPAS

	MBTS (n = 9)	RVPAS (N = 9)	
Patient characteristics			
Age (years)	5.3 ± 3.3	5.2 ± 3.4	NS
Stage II, n	3	3	NS
Stage III, n	6	6	NS
Male sex, n (%)	6 (67%)	8 (89%)	NS
Weight (kg)	17.3 ± 4.9	17.5 ± 6.5	NS
Cardiac magnetic resonance imaging analysis			
RVEDVi (mL/m^2)	97.2 ± 23.8	117.5 ± 26.6	NS
RVESVi (mL/m^2)	49.2 ± 16.8	67.0 ± 21.0	NS
RVEF (%)	50.0 ± 7.6	43.9 ± 5.4	NS
RV global mean peak circumferential strain (%)	−18.5 ± 4.8	− 13.9 ± 3.2	NS
RV global mean peak radial strain (%)	37.1 ± 15.1	25.0 ± 7.4	NS
RV global mean peak longitudinal strain (%)	− 16.0 ± 5.0	14.5 ± 2.8	NS

Data are presented as mean values. P values <0.05 are considered significant. legend: HLHS, hypoplastic left heart syndrome; MBTS, modified blalock-taussig shunt; RV, right ventricle; RVEDVi, indexed right ventricular end-diastolic volume; RVESVi, indexed right ventricular end systolic volume; RVEF, right ventricular ejection fraction; RVPAS, right ventricle-to-pulmonary artery shunt.

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at the presumed previous ventriculotomy site. Aneurysm orifice size measured 5–27 mm (median, 9 mm), and was not related to time interval since RVPAS (p = NS). Late gadolinium enhancement was present near the aneurysm in 7/20 (35%). Strain analysis by cMRI showed regional abnormalities at the aneurysm site as expected, with regional abnormalities extending to adjacent myocardial segments. The aneurysm was prospectively noted by TTE in only 1 subject. Nine age and stage matched RVPAS and MBTS subjects were compared (Table). MBTS group tended to have less RV enlargement and greater values of EF/strain, without reaching statistical significance.
Conclusion: HLHS patients with prior RVPAS universally had a dyskinetic aneurysm of the RV anterior wall. This defect may provide a nidus for ventricular arrhythmia, thrombosis and/or dysfunction in the long-term. Further research including cMRI analysis with larger number of subjects is needed.

P2925 - CARDIAC EVALUATION IN INFANTS WITH CONGENITAL ZIKA SYNDROME

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An outbreak of Zika virus (ZIKV) infection associated with congenital microcephaly has been reported in Brazil, especially in Northeast region. Prenatal Zica virus infection has been linked to adverse pregnancy and birth outcomes, most notably microcephaly and other serious brain anomalies. For these reasons, besides neurological defects, other abnormalities as cardiac involvement should also be excluded in infants with congenital Zika syndrome. To determine whether Zica virus infection during pregnancy causes cardiac malformation we performed echocardiogram in 113 patients diagnosed with Zica virus infection from November 2015 to May 2016. The patients were forwarded from the ambulatory independent of symptoms. From 113 patients, 61 were female. In 69 patients we observed some alterations considered as normal for the age group, as patent foramen oval, patent ductus arteriosus and small atrial septal defect. Only in 3 cases were diagnosed with ventricular septal defect, witch corresponds to 2,6% of the cases. This is in agreement with the findings of the literature in normal population. A few reports have described congenital malformation linked to Zika virus infection most of them in central nervous system. We found no association with congenital heart disease and Zika virus infection. Other studies are necessary to confirm this.

P2931 - VENTRICULAR FUNCTION ANALYSIS AT EXERCISE AS MEASURED BY REAL TIME CARDIAC MAGNETIC RESONANCE IMPROVES PREDICTION OF METABOLIC EXERCISE PERFORMANCE IN FONTAN PATIENTS

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Background: We wished to study the relationship between ventricular performance and exercise capacity in Fontan patients using a cardiac magnetic resonance (CMR) exercise protocol.
Methods: Fontan patients underwent standard graded metabolic exercise testing (MET) to evaluate metrics at peak exercise and anaerobic threshold (AT). A subsequent resting CMR included real-time (RT) cine imaging to quantify end-diasotlic (EDV), end-systolic (ESV), and stroke (SV) volumes, ejection fraction (EF) and indexed ventricular output (CI). Patients then underwent a graded exercise protocol on a supine ergometer to achieve the heart rate associated with AT on prior MET. Real-time measurements were repeated immediately after exercise. Flows were indexed to BSA and volumes to $BSA^{1.3}$ as previously described. VO2 at AT and peak Ex were indexed to body weight and compared with resting and exercise volumetric data using Pearson correlation. Significant factors were analyzed with multiple regression analysis.
Results: 46 patients age 16 years (range 12-42) had a mean EF of 65% (42-85%) at rest and 71% (53-87%) at exercise ($p < 0.001$). There was significant correlation between VO2 at AT and both resting CI ($r = 0.45$, $p = 0.001$) and change in CI from rest to exercise ($r = 0.34$, $p = 0.02$). In multiple regression analysis, both resting CI and change in CI remained significant. Other significant correlations with VO2 included both resting and ex SV, and ex CI. There was no significant correlation between EF, EDV, or ESV at rest or Ex with any MET parameter.
Conclusions: MET performance correlates with CI at rest, as well as change in CI from rest to exercise. Change in CI from rest to exercise improves the prediction of exercise performance compared to resting data alone. Characterizing ventricular function at exercise may yield a better understanding of how ventricular performance affects exercise capacity in Fontan patients.

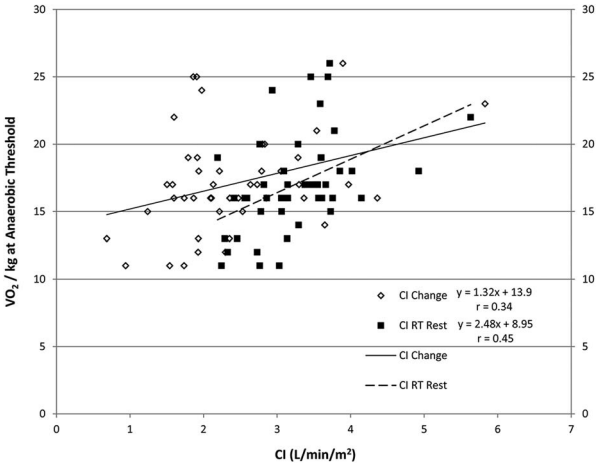

Figure.

P2936 - IMPACT OF PRENATAL DIAGNOSIS OF D TRANSPOSITION OF THE GREAT ARTERIES IN THE NEWBORN WHO REQUIRES A BALLOON ATRIAL SEPTOSTOMY

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Background/Hypothesis: It has always been assumed that a prenatal diagnosis of critical congenital heart disease would significantly improve postnatal outcomes by providing time to appropriately plan for management at delivery. However, for some conditions, the data has not been consistently supportive. We sought to investigate the clinical impact of a prenatal diagnosis in comparison to a postnatal diagnosis of simple D-transposition of the great arteries (DTGA) in newborns requiring urgent balloon atrial septostomy (BAS).

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Materials and Methods: We identified all newborns with simple DTGA (+/- small ventricular septal defect) who required BAS encountered in the province of Alberta between 2003 and 2015. Newborns delivered at <34 weeks and those with more complex congenital heart disease were excluded. We reviewed their postnatal charts and evaluated parameters indicative of their clinical status prior to BAS.
Results: We included 19 newborns with a prenatal diagnosis and 31 newborns with a postnatal diagnosis of simple DTGA. All infants were intubated and ventilated for BAS. Prior to BAS, postnatally diagnosed newborns had a significantly lower pH and higher peak arterial lactate compared to those with a prenatal diagnosis. Inotrope support was needed for 15% of those with a prenatal diagnosis versus 36% of those with a postnatal diagnosis. Nitric oxide was not required in any patients with a prenatal diagnosis, but 4/28 (14%) with a postnatal diagnosis. No patient required CPR or greater support and all patients survived to surgery. Median length of hospital stay was 18 days (range 9-45 days) for prenatally diagnosed and 21 days (range 11-92 days) for postnatally diagnosed groups ($p = 0.11$).
Conclusion: Prenatal diagnosis of DTGA significantly improves the preoperative clinical status of newborns requiring urgent BAS. The impact of improved preoperative condition secondary to prenatal diagnosis on enhancing long-term outcomes is the subject of ongoing investigations.

P2940-10 YEAR REVIEW OF PEDIATRIC CARDIAC MRI WITH DELAYED GADOLINIUM ENHANCEMENT; IMPACT ON OUTCOME

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Background/Hypothesis: In adults, the presence of delayed enhancement (DE) on cardiac magnetic resonance imaging (CMRI) is a useful modality in diagnosis and risk stratification. These practices have been applied in pediatrics at times without rigorous study. A pediatric CMRI study is not an easy undertaking due to requirements of patient compliance with need for general anesthetic for quality studies.
Our aim is to determine the types of patients currently undergoing DE on CMRI and to define the number of positive DE studies with a view to associate outcomes in our next study in order to better define which populations of cardiac patients truly benefit from the information acquired.
Materials & Methods: A retrospective review of all CMRIs performed at CHEO from September 2006-January 2016 and associated patient charts.
Results: At this time, our preliminary data shows there were 835 studies performed. In 50% (424/835) delayed enhancement was requested; 0.2% (2/835) studies could not be analyzed. Of the 424 patients, 10% (44/424) DGE was positive, 72% (305/424) DGE was negative, 5% (23/424) DGE could not be performed due to patient or technical factors, 11% (49/424) DGE was performed but not reported, and in 0.7% (3/424) studies there was either artifact or suboptimal imaging. The most common indications for studies were: ARVD in 28% (120/424) patients, TOF in 12% (50/424), and TGA or DORV in 16% (70/424). The male: female ratio was 1:6:1 with 516/838 males and 322/838 females receiving studies.
Conclusions: Over the last 10-year period, the observed finding of positive delayed enhancement in pediatrics was 10%. Our next study will determine the impact on outcomes.

P2941 - IMAGING OF THE DUCTUS ARTERIOSUS NORMAL ANATOMY PHYSIOLOGY AND CONGENITAL ABNORMALITIES

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Introduction: Ductus arteriosus [DA] is an artery that usually extends from the aortic isthmus, just distal to the left subclavian artery to the junction of the main and left pulmonary artery in fetal life. Knowledge of anatomy and physiology of DA is critical in management of a wide spectrum of congenital heart disease. Previously, variations in size and shape of the left sided DA have been well described. We aimed to describe variations in course, flow pattern, number, caliber in various patho-physiological conditions.
Methods: Our Radiology database of CT angiograms of the chest and cardiovascular MRIs, performed between January 1, 2008 and December 31, 2016, was interrogated for presence of DA. A total of 89 DA were reported, out of which 70 were beyond the first year of life. Descriptive text will be used to describe the features of the data in a study. We will discuss and illustrate the echo, CT and MR appearance of several clinically significant variations of the DA and discuss anomalies of the DA in terms of number, site, shape, course and size. Additionally, we will suggest techniques for CT and MR imaging of the DA with emphasis on determining their hemodynamic significance and importance in pre-procedural planning.
Findings: Variations in course of DA can be classified into -Vertical, reverse orientated, tortuous, cervical, right sided and bilateral. Other abnormalities of DA include enlargement of the DA, Abnormal flow in the DA, Absence of DA, DA aneurysm, thrombus in DA and endarteritis of DA.
Conclusion: CT and MR imaging provide excellent noninvasive depiction of the anatomy, flow and anomalies of the DA. Abnormalities of the DA range from numerical and morphologic variations to clinically significant anomalies. It can be important to identify these variations, to ensure appropriate treatment of congenital heart disease.

P2944 - CARDIAC FUNCTION ASSOCIATED WITH HOME VENTILATOR CARE IN DUCHENNE MUSCULAR DYSTROPHY

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Background: Duchenne muscular dystrophy (DMD) is prevalent neuromuscular disorder that affects one in 3600 live male births. Most leading cause of mortality in DMD patients is usually respiratory complications or cardiomyopathy. But nowadays, cardiomyopathy is becoming the leading cause of death in DMD patients because mechanically assisted lung ventilation and assisted coughing help resolving respiratory complication. We investigated to reveal the relation of cardio-pulmonary function in DMD patients.
Method: We retrospectively reviewed the DMD patients diagnosed from 2010 January to 2016 March in Gangnam Severance Hospital. Demographic characteristics, pulmonary function factor, Echocardiography data were assessed.
Results: We reviewed 54 case of DMD and divided into two groups (Home-ventilator assisted group vs Non-ventilator assisted group). In pulmonary function analysis, Home-ventilator group (1038 ± 620.41 ml) show lower sitting Functional vital capacity than Non-ventilator group (1455 ± 603.12 ml). In echocardiographic measurement, mean LV Ejection fraction and Fractional shortening were higher in Home-ventilator group. Home-ventilator

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assisted group's E/A ratio(1.7 ± 0.44) was more stable than Non-ventilator assisted group(2.02 ± 0.62). With estimation by the multiple linear regression test, TDI S' was higher in Home-ventilator group (Estimated β : 1.06, Standard error : 0.48).
Conclusion: DMD patients with ventilator care might have better systolic and diastolic cardiac function. Especially, patients without ventilator assistance may need more meticulous evaluation for cardiopulmonary function, which may help to care the early cardiopulmonary dysfunction.

P2947 - CORONARY ARTERY LESIONS IN COSTA RICAN CHILDREN WITH KAWASAKI DISEASE A 5 YEAR RETROSPECTIVE STUDY (PERIOD 2009 2013)

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Background/Objectives: Kawasaki disease (KD) is the leading cause of acquired heart disease in Costa Rican (CR) children (ch). Despite its importance, no recent analysis focusing on children who develop coronary artery lesions (CALs) has been made at our institution. We describe the epidemiology, clinical aspects and treatment of patients (pts) with KD who developed CALs.
Materials and Methods: Retrospective descriptive study of hospitalized pts with a KD diagnosis, who developed CALs and were attended at the only national pediatric tertiary referral teaching hospital of CR. Study period: Jan-1st-2009 to December-31st-2013.
Results: Among 149 enrolled pts with KD, a baseline echocardiogram (ECHO) was performed in all. CALs were detected in 15 (10.1%) pts as follows: dilation, 8 pts (53.3%); aneurysms, 7 (46.7%) pts. Incomplete KD occurred in 3 (20%) pts, all of which developed coronary aneurysms. All pts were hospitalized; 9 (60%) pts were male. Mean age at admission was 32 (3-92) months, distribution by age groups was: <6 months, 3 (20%) pts; 7-24 m, 6 (40%) pts; 25-60 m, 3 (20%), and >60 m, 3 (20%) pts. Mean length of fever at admission was 13 (3-30) days. Prior to admission and KD diagnosis, all 19 pts had at least >1 recent medical visit and all pts received antibiotics for other presumed diagnoses. IVIG treatment was administered in 13 (86.7%) pts: 1 dose, 9 (69.2%), and 2 doses, 4 (30.8%), respectively. Aspirin and steroids were given in 19 (100%) and 1 (6.7%) pts, respectively. No deaths occurred.
Conclusions: Among children with CAL's, the rate of IVIG resistance is higher than other reports from LA. The high rate of antibiotic use prior to final KD diagnosis suggests that clinical suspicion of the disease is low.

P2965 - USEFULNESS OF MYOCARDIAL PARAMETRIC IMAGING TO EVALUATE PHYSIOLOGICAL CHANGING IN LEFT VENTRICULAR NONCOMPACTION ASSOCIATED VIRAL MYOCARDITIS BEFORE AND AFTER REDUCING VOLUME OVERLOAD

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The left ventricular sponge morphology fulfilling the diagnostic criteria for left ventricular noncompaction (LVNC) may occur in association with acute deterioration in left ventricular function of various causes. This may occur as consequence an adaptive mechanism aimed at increasing endocardial surface area and optimizing stroke volume. The combination of viral myocarditis and LVNC become dillema piffall missdiagnosis of main caused left ventricular function depress. We report the case of a 1 year old girl with diagnosis patent ductus arteriosus (PDA) presented heart failure symptoms and viral myocarditis infection. Three dimensional echocardiography (3DE) accidently finding LVNC morphology. The patient was evaluated by 3DE parametric imaging , Mitral Valve (MV) inflow, Tissue doppler imaging (TDI) for before, intermediate, after 1 day and after 3 day PDA occlussion procedure. This evaluation study to define the changing in left ventricular (LV) delay contraction, myocardium contraction and MV inflow after reducing left ventricular volume overload by closing PDA shunt. The result; End diastolic LV volume reducing from 43.5 ml into 36.6 ml and LV ejection fraction improved from 26% to 44%. The MV inflow has showed significant change from absent A wave into normal patent of MV inflow. The parametric imaging of bull eye has demonstrated signicant change timing delay contraction and myocardium contraction except basal and apex region. Conclusion; The use of 3DE parametric imaging allowed evaluation of hemodynamic change based echocardiography.

P2974 - PRENATAL FINDINGS IN HYPOPLASTIC LEFT HEART SYNDROME

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Background/Hypothesis: Hypoplastic left heart syndrome (HLHS) is one of the most common congenital heart defects detected prenatally. The aim of this study was to describe the prenatal findings in a cohort of patients with HLHS.
Materials And Methods: Retrospective descriptive study. Clinical records and fetal echocardiographic clips of patients with HLHS diagnosed prenatally in 2 tertiary health care centers in a 5-year period were reviewed.
Results: Sixty seven patients with prenatal diagnosis of HLHS were assessed in 2 tertiary health care centers, with median maternal age of 35 years old (range 17 to 42 years old) and median gestational age at initial assessment of 33 weeks (range 21 weeks 6 days to 38 weeks 3 days). Five/67 (7.5%) cases had associated major extracardiac and/or genetic anomalies. There were 35/67 (52.2%) fetuses with aortic and mitral atresia, 14/67 (20.9%) with aortic atresia and mitral stenosis, and 18/67 (26.9%) with aortic and mitral stenosis. Eight/67 (11.9%) patients had a severely restrictive or intact atrial septum, 3/67 (4.5%) severe tricuspid regurgitation, and 2/67(3%) right ventricular dysfunction. The median of right/left ventricular ratio was 2.14 (range 1 to 14.4), of pulmonary/aortic artery ratio was 2.2 (range 1,18 to 5,33), and of pulmonary /aortic annulus ratio was 2,55 (range 1,3 to 5,5). There were 12/67 (17.9%) patients lost to follow up, 4/67 (6%) received comfort care and 51/67 (76.1%) underwent multisage palliative surgery.
Conclusion: The prenatal findings in a cohort of patients with HLHS are described. In this cohort a low association with

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extracardiac and genetic anomalies was observed, and most patients underwent multisage palliative surgery.

P2977 - HYPOPLASTIC LEFT HEART SYNDROME EARLY MORTALITY

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Background/Hypothesis: Survival of patients with hypoplastic left heart syndrome (HLHS) has steadily improved since the advent of the multistage reconstructive approach for surgical palliation. Poor outcome has been associated to patients´ high-risk factors. The aim of this study was to examine early survival in a cohort of patients with HLHS, assessing the impact of high-risk factors.
Material and Methods: Clinical records of patients born in a 5-year period with HLHS in 2 tertiary specialized hospitals were reviewed. Early survival was evaluated in relation to the presence of 1 or more of the following high-risk factors: major extracardiac, genetic or chromosomal anomalies; prematurity (<34 weeks of gestation at birth); low birth weight (<2500grs); intact or highly restrictive atrial septum; severe tricuspid regurgitation, or severe ventricular dysfunction. Differences in survival were assessed using contingency table and Fisher exact test analysis.
Results: Of 62 patients 55 (88.7%) had prenatal diagnosis, there was 1 premature and 3 low birth-weight patients. There were major extracardiac malformations and/or genetic disorders in 5/62 (8.1%) patients. There was intact or highly restrictive atrial septum in 8/62 (12.9%), severe tricuspid regurgitation in 2/62 (3.2%) and right ventricular dysfunction in 1/62 (1.6%). Among the 17/62 (27.4%) patients with 1 or more risk factors: 3 received comfort care, 3 died before surgery, and 5 died after Norwood surgery (57.1% early mortality). Forty five/62 (72.6%) patients did not have risk factors, and 2 of them died before surgery and 13 after Norwood surgery (33.3% early mortality). Early mortality was increased in the group with high-risk factors (p 0.0423).
Conclusions: Nearly one third of patients with HLHS had associated high-risk factors, which increased their early mortality. Identifying these risk-factors in the perinatal period is important for assessing prognosis in patients with HLHS.

P2979 - FETAL AORTIC VALVULOPLASTY

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Background/Hypothesis: Fetal aortic valvuloplasty has been proposed for preventing the progression to hypoplastic left heart syndrome (HLHS) in patients with critical aortic stenosis and specific echocardiographic criteria. The aim of this study is to report the experience of our group in this disease.
Materials and Methods: The medical records of patients with critical aortic stenosis diagnosed prenatally between February 2005 and February 2016 were reviewed. Measures of central tendency and frequency distribution were calculated.
Results: Critical aortic stenosis at risk of progression to HLHS was found in 16 cases, and 14/16 (87.5%) underwent fetal aortic valvuloplasty at a median gestational age of 27 weeks (range 20.1 to 30.5 weeks), with technical success in 12/14 (85.7%). In 4/14 (28.6%) fetuses death occurred within 48 hours of the intervention. At birth HLHS was observed in 2 cases that had not been intervened prenatally, in one patient with technical failure and in 1 of those who underwent fetal aortic valvuloplasty. Regarding patients with technically successful procedures, 7/8 patients had biventricular physiology and left ventricles with preserved dimensions or borderline cases. No significant maternal complications were observed.
Conclusions: Fetal aortic valvuloplasty is feasible, without significant maternal risks, to be performed by trained intervention teams in selected patients. The results so far are encouraging, but given the complexity of the pathology clinical impact is still under study.

P3014 - LEFT VENTRICULAR MYOCARDIAL REMODELING IN FOETAL LIFE

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Background: Left ventricular myocardium is a strongly anisotropic tissue in adults. Cardiomyocytes are arranged in a complex three-dimensional network which forms a counterclockwise helix in the endocardial layer and a clockwise helix in the epicardial layer. This peculiar organization is fundamental to myocardial functionality because left ventricular contraction is not only longitudinally and radially oriented, but also characterized by ventricular twisting. Foetal physiological myocardial development dynamics has not been completely explained yet and the few studies about this topic focused mainly on the histology of mammalian hearts and not human ones.
Aim of the study: Speckle Tracking Echocardiography allows to study separately endocardial and epicardial functionality in vivo. Combining this data with morphometrical observation on histological sections of fetal human heart, we compared the development of the different myocardial layers with advancing gestational age.
Methods: To study foetal myocardial function, we enrolled 72 foetuses and 39 premature babies without cardiac pathologies. We studied longitudinal endocardial and epicardial strain in these patients. For the histological section, we studied 20 foetal hearts without cardiac pathologies from autoptical investigation. We determined every layer's thickness and cardiac fibres orientation in comparison with gestational age.
Results: Endocardial values of deformation are higher than epicardial ones. Epi/endocardial values ratio increase with gestational age (figure). The epicardial layer, in fact, becomes more influencing. Histological data confirm our observations. All the layers increase their thickness during gestation but epicardium increases fourfold while trabecular and endocardial layers maintain the same value throughout pregnancy.
Conclusions: Left ventricular myocardium maturation is a process that begins early during fetal life. It starts from the differentiation of a subendocardial layer. The development of the epicardial is slower and is completed late in the foetal life. Probably this process is led by the increasing pre- and after-load in the third trimester of gestation.

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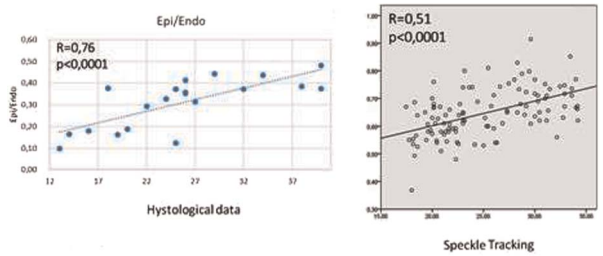

Figure.

P3016 - CAROTID SUBCLAVIAN ARTERY INDEX IMPORTANT TWO DIMENSIONAL ECHOCARDIOGRAPHIC PARAMETER FOR DETECTION OF COARCTATION OF AORTA IN NEONATES

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Background: Coarctation of aorta is a very common congenital heart malformation occurring in 6-8% of all congenital heart diseases. However in neonates coarctation may be missed or underestimated by echocardiography, especially with patent ductus arteriosus. The carotid-subclavian artery index: the ratio of the aortic arch diameter at the left subclavian artey to the distance between left carotid artery and the left subclavian artery, has been proposed in previous studies for establishing the diagnosis of coarctation of the aorta.
Objective: To evaluate such indexes and to look for other echocardiographic predicators of coarctation of the aorta. *Design/Methods:* Echocardiographic evaluations was retrospectively reviewed in patients with coractation of the aorta as well as control group admitted at a tertiary cardiac center in the period between January 2010 to December 2014. All patients admitted with coarctation of aorta either had surgery or catheter intervention which confirmed the diagnosis. 39 patients with Coarctation as well as 20 patients in controls were included in the study. End systolic measurements were obtained from 6 different sites of the aortic arch.
Results: The distance between the origins of the great vessels: Brachiochepahic to Left carotid artery (median 2.8 mm vs 1.5 mm in controls) and Left carotid artery to Left Subclavian artery (median 6.22mm vs 2.3mm in controls) were longer in patients with coarctation than in controls. Carotid-subclavian artery index was significantly smaller (Median 0.59, Range 0.28-2.67) in the coractation group in comparison to the control group (Median 2.8, Range 1.56 -4). 92% patients with coarctation had a Carotid-subclavian artery index <1.
Conclusion(s): A carotid-subclavian artery index of <1 is a useful noninvasive screening parameter for coarctation and may be useful in unstable patients or in those with a patent ductus artersious in which coarctation may be overlooked.

P3017 - TRADICIONAL AND ADVANCED PARAMETERS OF CARDIAC MECHANICS IN CHILDREN WITH END STAGE RENAL DISEASE AT HOSPITAL INFANTIL DE MÉXICO FEDERICO GÓMEZ

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Background and Objectives: Cardiovascular disease is recognized as a major cause of death in children with end-stage renal disease (ESRD). The aim of this study was to identify prevalence of abnormalities in left ventricular function through the analysis of tradicional and advanced indices of cardiac mechanics in children with ESRD.
Methods: Between 2015 and 2016, the cross sectional, prospective and observational study enrolled children with ESRF ages 1–17 years old undergoing hemodialysis. Cardiac morphology and function were assessed through transthoracic echocardiography. The analysis presented encompasses global radial, longitudinal, circumferential and torsión strain. The patients were divided en two groups: from 1 to 9 years (group 1) and from 10 to 17 years (group 2). Data were matched with children with comparable age and sex.
Results: Data on 47 patients with ESRF with complete echocardiographic assessment are reported (age = 11.1 ±4.8 years). Patients with ESRD showed mildly higher office Blood Pressure values and higher prevalence of left ventricular hypertrophy, but no differences were observed among groups in left ventricular ejection fraction. In group 1, strain analysis showed significantly lower epicardial (-14.75 ± 3.58 vs -17.7 ± 3.71 p 0.05) and endocardial strain (-19.04 ± 3.77 vs -22.52 ± 4.23 p 0.03) in patients with ESRD, resulting in a significant difference in global longitudinal strain (-16.25 ± 3.00 vs -19.01 ±3.7 p 0.03). In group 2, Strain analysis showed significantly lower global radial strain in patients with ESRD (25.72 ± 9.53 vs 33.98 ± 14.16 p 0.006) without differences among groups in global radial, circunferencial and torsion strain.
Conclusions: A significant proportion of children with ESRD undergoing hemodialysis show impaired systolic mechanics. In younger patients, impaired systolic function is characterized by lower endocardial and epicardial longitudinal strain. In teenagers impaired systolic function characterized by lower global radial strain.

P3041 - MITRAL REGURGITATION CAUSED BY A CLEFT IN THE POSTERIOR MITRAL VALVE LEAFLET. CASE REPORT

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The cleft in the posterior mitral valve leaflet is a very rare congenital anomaly that causes mitral regurgitation. Most of the clefts involve the anterior leaflet and isolated clefts of the posterior leaflet have been anecdotally identified.
Case 1: We report the case of a 17 years old female, with Down syndrome. She was referred for evaluation of a cardiac murmur. Auscultation revealed a systolic murmur at high parasternal border. Transthoracic two-dimensional and transesophageal three-dimensional echocardiography showed moderate mitral regurgitation originated through the cleft of the posterior valve.

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In addition, a prolapse of the anterior mitral leaflet was observed, contributing with the regurgitation. Additionally we found an atrial septal defect.

Case 2: A ten years old female with Marfan syndrome. She was referred for evaluation of a cardiac murmur and as part of Marfan syndrome evaluation. She was asymptomatic and auscultation revealed a systolic murmur at midline fifth intercostal space. Transthoracic two and three dimensional echocardiography showed 2 jets of moderate mitral regurgitation caused by a cleft in the posterior mitral valve leaflet and a prolapse of the anterior mitral valve (A2 scallop), also we found a tricuspid septal leaflet prolapse with mild regurgitation, an atrial septal defect and a dilated aortic root.

In summary, in mitral regurgitation caused by a cleft in the posterior mitral valve the severity of regurgitation is variable. We found just one case reported in a child previously and the cases reported here are even rare because of its occurrence with Marfan and Down Syndromes.

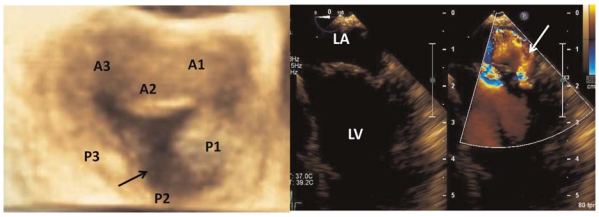

Figure 1.
Patient 2, transesophageal echocardiogram three dimensional image of the cleft in the posterior mitral valve leaflet and two dimensional color of mitral regurgitation. Black arrow shows the cleft, white arrow sohws the mitral regurgitation. AL: anterior leaflet of the mitral valve; PL: posterior leaflet of the mitral valve LV: left ventricle, LA: left atrium.

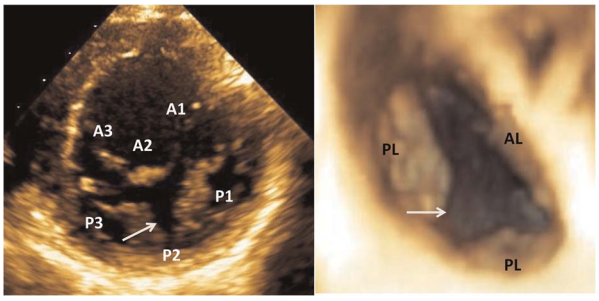

Figure 2.
Patient 2, two and three dimensional image of the cleft in the posterior mitral valve leaflet. White arrow shows the cleft. AL: anterior leaflet of the mitral valve; PL: posterior leaflet of the mitral valve

INTENSIVE CARE

P1011 - A NEW LOOK AT BRONCHOPULMONARY DYSPLASIA POST CAPILLARY PATHOPHYSIOLOGY AND CARDIAC DYSFUNCTION

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Background: In preterm infants, pulmonary hypertension (PH) and right ventricular function are the focus of cardiovascular effects of bronchopulmonary dysplasia (BPD). The impact of left sided (systemic) indices is not well understood.

Objective: To assess cardiac indices reflecting systemic afterload and pulmonary venous back pressure as pathophysiologic factors in infants with 'severe' BPD.

Methods: Cardiac parameters were measured by conventional echocardiography in 20 preterm infants with severe BPD and compared with 10 preterm infants with no BPD and 20 healthy term infants.

Results: The gestational age and birthweight amongst preterm infants with and without BPD were comparable (26.2 ± 1.7 vs 26.2 ± 0.6 weeks and 772 ± 271 vs 704 ± 53 g). All the infants belonged to the 'severe BPD' category (need for $\geq$ 30% oxygen and/or positive pressure at 36 weeks post menstrual age). None of them were intubated and mechanically ventilated at the time of the study. In infants with severe BPD, PH was noted in [5 (25%), tricuspid regurgitation Doppler jet ≥ 2.8 m/s and 15 (75%), by the Time to peak velocity/Right ventricular ejection time < 0.34]. Amongst systemic cardiac indices, significant impairment of diastolic function was noted in the BPD group compared to no BPD infants and term infants. The significance persisted after adjusting for gestational age and birthweight. A higher end systolic wall stress (surrogate for afterload) had a significant correlation with lower mean velocity of circumferential fibre shortening (contractility) ($r = -0.74$, $p = 0.0002$).

Conclusions: Altered systemic (left sided) cardiac function was noted in the infants with BPD which may lead to pulmonary venous congestion contributing to a continued need for respiratory support. Impaired left ventricular relaxation and high left atrial pressure may be oedemagenic factors.

Table 1. Comparison of echocardiographic parameters

Variable	BPD group (n = 20)	Preterm no BPD group (n = 10)	Term infants (n = 20)	Unadjusted overall group effect (p value)	GA adjusted group effect (p value)	BW adjusted group effect (p value)	GA and BW adjusted group effect (p value)
Heart rate (beats/min)	146 ± 10	152 ± 11	145 ± 8	0.24	0.2	0.13	0.44
Diastolic parameters							
E wave deceleration time (ms)	75.6 ± 7.9	63.6 ± 8.2	60.5 ± 7	<0.0001	0.001	0.001	0.001*
End systolic wall stress (g/cm2)	72 ± 10.8	44.2 ± 6.5	41.8 ± 6.4	<0.0001	<0.0001	<0.0001	<0.0001*
Trans-mitral E/A	1.07 ± 0.07	0.91 ± 0.04	0.89 ± 0.09	<0.0001	<0.0001	<0.0001	<0.0001*
Iso volumic relaxation time (ms)	68.8 ± 3.9	58.5 ± 7.8	54.2 ± 5.7	<0.0001	<0.0001	<0.0001	<0.0001*
Mitral valve stroke volume (ml/kg)	4.7 ± 0.7	5.6 ± 0.6	5.9 ± 0.1	<0.0001	0.01	<0.0001	0.002*
Pulmonary vein VTI (cm)	5.4 ± 1.8	7.8 ± 0.9	8.2 ± 1.3	<0.0001	<0.0001	<0.0001	<0.0001*
Tei index	0.33 ± 0.05	0.28 ± 0.01	0.27 ± 0.05	0.001	0.03	0.004	0.03*
Systolic parameters							
Mean velocity of circumferential fibre shortening (circ/s)	1.7 ± 0.4	2.7 ± 0.2	3 ± 0.6	<0.0001	<0.0001	<0.0001	<0.0001*
Left ventricle stroke volume (ml/kg)	1.25 ± 0.32	1.29 ± 0.09	1.32 ± 0.21	0.72	0.03	0.48	0.046
Left ventricular output (ml/kg/min)	183 ± 45	189 ± 9	191 ± 32	0.75	0.054	0.21	0.03
Fractional shortening (%)	33.1 ± 3.3	33.3 ± 2.8	33.5 ± 0.5	0.96	0.95	0.93	0.92

Data are presented as mean ± standard deviation. * BPD Group vs No BPD Group and BPD Group vs Term controls are significant ($p < 0.05$).

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P1013 - A LONG TERM FOLLOW UP OF AN EXTREMELY LOW BIRTH WEIGHT INFANT WITH RIGHT ISOMERISM AND MULTIPLE CONGENITAL HEART ANOMALIES

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Background: The prognosis of right isomerism heart with total anomalous pulmonary vein connection is still not satisfactory. And complex congenital heart disease in extremely low birth weight patient is also not favorable.
Case: We experience an extremely low birth weight infant, born 29 weeks 4 days and birth weight was 879 g. She also had the right isomerism heart with complete atrioventricular septal defect, double outlet right ventricle, pulmonary atresia, and patent ductus arteriosus. Initially, pulmonary blood flow was controlled by using intermittent prostaglandin infusion. At the age of 5 months, left modified Blalock-Taussig shunt was done. Bidirectional Glenn shunt was performed at the age of 12 months old. And finally total cavo-pulmonary connection was done at the age of 24 months. After Fontan operation, hemodynamic status remained stable with using some anti-failure medications, but she shows mild pulmonary vein stenosis at the recent follow-up examination. We are planning to do the catheter examination. She suffered severe streptococcus pneumoniae sepsis when she was 2 years 10 months old. Other than that, no critical infection was issued till today. Regarding her development, she is slow in learning. She is now 10 years old but her emotional development is 6 years old and language development is 7 years. Now she is going to special supportive class in the municipal elementary school.
Conclusion: Prognosis of an extremely low birth weight patient with complex heart anomalies is not known. No case reports are found by PubMed search. Our case shows satisfactory clinical course. Aggressive treatment for this patient group may be acceptable.

P1017 - VASOACTIVE VENTILATION RENAL SCORE AND ICU LOS IN PATIENTS UNDERGOING RE ENTRY STERNOTOMY FOR CONGENITAL HEART DISEASE A SINGLE INSTITUTE EXPERIENCE

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Purpose: Severity of injury scores have not reliably predicted ICU LOS in patients undergoing surgery for congenital heart disease (CHD.) In those patients undergoing re-entry sternotomy, limited information regarding outcomes is available. A newer score, the Vasoactive-Ventilation-Renal Score (VVR score) may be used to predict ICU LOS after re-entry sternotomy for CHD.
Methods: Retrospective analysis of patients undergoing reentry sternotomy for CHD at our center between August 1, 2009 and May 31, 2016 was performed. Vasoactive-Ventilation-Renal (VVR) scores were calculated at admission post-procedure, 24-hours after admission, and at 48-hours. We evaluated injury of structures on entry, cardiopulmonary bypass time, cross-clamp time, blood requirements within 48 hours, duration of mechanical ventilation, ICU and hospital LOS, low cardiac output syndrome, sepsis, bleeding, renal failure, arrhythmias, and death. The response variable was ICU length of stay in days. Recursive partition analysis was used to identify the variables predicting length of stay. Of the initial 25 features, 5 were removed for near zero variance and 3 categorical features were removed for non-information. Covariance analysis did not demonstrate any significant correlation among the remaining features. Initial recursive tree regression using ANOVA, cross validation and cp = 0.01 produced 4 trees. To avoid over-fitting, the tree with lowest cross validation error was selected.
Conclusions: The resulting 2 split trees identified ventilator days and VVR at 48 hours to be predictive of ICU LOS.

P1018 - A RETROSPECTIVE COHORT STUDY TO EVALUATE THE USE OF PARACETAMOL COMPARED TO IBUPROFEN IN THE MEDICAL CLOSURE OF PATENT DUCTUS ARTERIOSUS AMONG PRETERM INFANTS IN A PRIVATE TERTIARY HOSPITAL

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Background: Persistent Patent Ductus Arteriosus (PDA) results from failure of the physiologic closure of the fetal ductus which renders the preterm newborn vulnerable to pulmonary overcirculation with diminished systemic blood flow. In this case, therapeutic intervention to facilitate ductal closure might be indicated. Objective: To determine the effectiveness and safety of oral and intravenous paracetamol compared to oral ibuprofen in the closure of a PDA in preterm infants.
Method: A retrospective cohort study was performed involving 28 cases of premature infants at a private tertiary hospital from January 1,2010 to June 30, 2014. Each patient received either oral paracetamol at a dose of 15 mg/kg every 6 hours for 3 days or oral ibuprofen at an initial dose of 10 mg/kg followed by 5 mg/kg at 24 and 48 hours.
Results: The PDA closure rates for both paracetamol and ibuprofen were identical at 64.3% (9/14). The mean days of closure for the paracetamol group was shorter at 1.5 ± .46 days in contrast to the ibuprofen group which is 2.7 ± .48 days (P = 0.00) There were no significant differences between the two groups in the incidence of adverse events. There were more preterms that had gastrointestinal bleeding in the ibuprofen group (28.6%) compared to the paracetamol group (7.1%) (P = 0.32) Platelet counts in the ibuprofen group decreased after one week of treatment at -87 ± 115.84, while that of paracetamol group increased by 12 ± 92.34 (P = 0.48)
Conclusions: Paracetamol may have a place with equal efficiency with Ibuprofen, the standard drug of choice, in the medical treatment for closure of PDA. Paracetamol with its lower incidence of adverse events may be a better alternative drug in the treatment of PDA in preterm infants.

P1040 - PAROXYSMALSUPRAVENTRICULAR TACHYCARDIA (PSVT) IN CHILDREN – THERAPEUTIC APPROACH IN URGENT PEDIATRICS

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Objectives: 1. Determination of trigger factors against the background of congenital heart defect. 2. Clinical manifestations of PSVT depending on age. 3. Effectiveness of treatment depending on age. Relevance of the topic: PSVT is one of major urgencies in children.

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Materials and Methods: The study included a group of 22 children and was based on survey and monitoring in dynamics in Emergency Receiving Pediatrics Unit Department, Clinical Hospital Balti in 2014.
Study results: 1. Age separation: 1st lot 0-4 years old (8 children – 36,36%), 2nd lot 5-18 years old (14 children – 63,63%) 2. Etiology separation: 1st lot PSVT with acute respiratory infection, pneumonia, congenital heart defect (atrial septal defect (ASD), ventricular septal defect (VSD)) – 36,36% 2nd lot PSVT in intact heart (stress, fear) – 63,63% 3. Clinical manifestation separation: 1st lot – cyanosis, extreme pallor, dyspnea – 36,36% 2nd lot – palpitation, heart pain, pallor, asthenia – 63,63% 4. Treatment separation: 1st lot – attack treatment with Adenosine – 36,36% 2nd lot – attack treatment with Amiodarone – 39,28%, Verapamil – 24,35%.
Conclusions: 1) High frequency of PSVT in children with concomitant diseases against the background of congenital heart defects (ASD, VSD) 2) High effectiveness of attack treatment in 1st lot with Adenosine, 2nd lot with Amiodarone and Verapamil 3) In Emergency Receiving Pediatrics Unit Department it is possible to treat PSVT attack in children.

P1065 - RISK FACTORS FOR THE DEVELOPMENT OF GIANT CORONARY ANEURYSMS IN THE ACUTE PHASE OF KAWASAKI DISEASE IN MEXICAN PATIENTS

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Background: An important complication of Kawasaki disease is the developement of giant coronary aneurysms. Risk factors for their development are not still clear.
Methods: A retrospective analysis was conducted at the Instituto Nacional de Pediatría in Mexico City, Mexico. It included all patients with diagnosis of Kawasaki disease between 1995 and 2015. Clinical and laboratory findings, as well as echocardiographic measurements were recorded. Patients with giant coronary aneurysms were used as the case group and all the other patients as a control group. P <0.05 was considered to be statistically significant. OR and their 95% CI were calculated to delinate the risk factors.
Results: 416 patients were diagnosed as Kawasaki disease from 1995 to 2015. 34 developed giant coronary aneurysms at the acute stage of the disease. In multivariate analyses, patients aged younger than 1 year (p <0.001 OR = 3.827 IC 95% 1.862-7.867) showed significantly higher OR for giant coronary aneurysms. In univariate analyses changes in the palms (p <0.028), the presence of congestive heart failure (p < 0.000472), or neurological findings (p < 0.01), as well as atypical manifestations (p <0.044) were found to significantly increase the risk of developing giant coronary aneurysms. Those who received additional intravenous immunoglobulin (p <0.023), and patients with higher duration of illness at time of presentation to hospital, showed significantly higher risk for giant coronary aneurysms. Anemia, leukocytosis, thrombocytosis, a lower erythrocyte sedimention rate and hipoalbuminemia were also found as risk factors. The presence of extracoronal findings on echocardiography (myocarditis, pericardial effusion, pericarditis and hypokinesia) were significantly associated with the development of giant coronary aneurysms.
Conclusions: One of the main factors associated with the development of giant coronary aneurysms was the delay in diagnosis of Kawasaki disease. This findings demonstrate the importance of maintaining a high suspicion of the disease, enabling early diagnosis and prompt.

P1066 - ACUTE MYOCARDIAL INFARCTION IN THE ACUTE PHASE IN KAWASAKI DISEASE IN MEXICAN CHILDREN

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Background: Kawasaki disease is an acute febrile vasculitis of unknown origin. Despite treatment with intravenous immunoglobulin during the acute phase of the disease, up to 5% of those affected will develop coronary aneurysms predisposing them to thrombotic complications that could result in myocardial infarction (AMI). In Mexico there are few reports of ischemic complications secondary to KD.
Objective: To describe the clinical features, the laboratory parameters, treatment used and the outcome of children who presented with myocardial infarction during the acute phase of KD in a third level facility in Mexico City.
Methods: From our Institutional Database of KD we search for children who presented AMI in the acute phase of the disease from August 1995 to August 2016. We analyzed gender, age, clinical manifestations, time from the onset of the symptoms to diagnosis, laboratory parameters, treatment used, and outcome in the acute phase of the disease.
Results: Eight infants were diagnosed with AMI during the study period. The median age at diagnosis was 8 months (range 2 to 53 months). Seven patients were male (87.5%). The median from the onset of the clinical manifestations to diagnosis of KD was 22 days (range 4 to 26 days). All patients developed giant coronary aneurysms (median Z-score 18.98, with a range of Z-score from 11.58-27.70). An abnormal EKG and abnormal perfusion tests demonstrated the myocardial infarction in all cases. Two patients died in the acute phase of cardiogenic shock, one more patient died of dilated cardiomyopathy 12 months after coronary bypass surgery with an overall mortality of 62.5% of this group.
Conclusions: AMI is a fatal complication of KD. In our small series it was associated with a delayed diagnosis of the disease and therefore the development of giant coronary aneurysms. Treatment of AMI in children after KD is a medical challenge.

P1093 - INTERSTAGE READMISSIONS IN SINGLE VENTRICLE PATIENTS UNDERGOING STAGED PALLIATION

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Background: Readmissions during the interstage period are common in infants with single ventricle congenital heart disease

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undergoing staged surgical palliation (S1P). We retrospectively examined readmissions during the interstage period.
Methods: Newborns undergoing S1P from January 2012-May 2015 who survived to hospital discharge were reviewed (n = 72). Patients followed elsewhere were excluded (n = 7). All of the patients undergoing S1P were home monitored.
Results: Study group comprised 65 patients. Surgeries included stage 1 hybrid palliation (44), Blalock-Taussig shunt (14), pulmonary artery band (7). Forty-nine patients had a total of 91 interstage readmissions; 86% were unscheduled. There were 19 major interstage adverse events (21%) - arrest/acute life threatening event (5); stroke (1); sepsis (1); cardiac catheterization requiring intervention - enlargement of atrial septal defect (3), retrograde aortic arch stenosis (2); evaluation for mesenteric ischemia (1); pneumonia/aspiration (2); shock (2); pericardial effusion requiring drainage (1); volvulous requiring bowel resection (1). Twenty-five readmissions (27%) were secondary to cardiac reasons defined as changes on echocardiogram - decreased function, restrictive atrial septal defect, retrograde aortic arch stenosis, stenotic BTS, or scheduled cardiac catheterization. Six patients suffered interstage deaths (9%). Nine patients (14%) spent >30 days in the hospital during the interstage period. Of the 55 newborns (85%) discharged following S1P 100% orally feeding; 73% remained 100% orally feeding throughout interstage period. No patient discharged with NGT/GT feedings learned to eat during the interstage period.
Conclusion: Interstage readmissions are common, with the majority being unscheduled. Interstage mortality was <10%. Majority of patients discharged 100% orally feeding following S1P remained 100% orally feeding throughout the interstage period. However, patients did not "learn to eat" during the interstage period.

P1130 - CAN ACUTE PHASE RESPONSE BIOMARKERS DIFFERENTIATE INFECTION FROM INFLAMMATION POST PEDIATRIC CARDIAC SURGERY

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Infection is common serious complication post pediatric cardiac surgery, diagnosis of infection is difficult in presence of surgical stress and inflammatory reaction after cardiopulmonary bypass.
Aim: The purpose of this study is to investigate the value of available inflammatory biomarkers and its validity in differentiate infection from inflammation post pediatric cardiac surgery. And find out the trend and the figures of these biomarkers after cardiac surgery.
Methods: We conducted a prospective study included all pediatric patient underwent cardiac surgery Erythrocyte Sedimentation Rate , White Cell Count with Neutrophile count pre surgery and for 4 consequence days post surgery and pre discharge were collected for all patients. Patients were divided into two groups infected and non infected group and off- on pump group then group who received corticosteroid pre surgery and others who didn't receive. Data were reviewed and analyzed.
Results: During study period 134 pediatric cardiac patients were collected . non infected group was 125 cases and infected group was 9 cases. We found No statistical difference in inflammatory biomarkers elevation between the groups the infected group had higher RACSH score and more days on ventilator and they had more drop in platelet count comparing with non infected group in day 2 and 3 post surgery. Both patients in on and off pump had same level of inflammatory biomarkers. Giving corticosteroid didn't change the trend of serum elevation of biomarkers.
Conclusion: Available inflammatory biomarkers cant differentiate between infection and inflammation during 5 days post cardiac surgery.

P1134 - EARLY VASCULAR AGING AND MALADAPTIVE ARTERIAL VENTRICULAR INTERACTIONS IN PRETERM INFANTS WITH FETAL GROWTH RESTRICTION

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Background: The cardiac adaptations already present in preterm FGR infants soon after birth are not known. The objectives of the study were to assess vascular and cardiac morphology and biomechanics in preterm infants with FGR compared to an appropriate for gestational age (AGA) cohort.
Methods: 20 preterm infants (28-32 weeks) GA and birthweight (BW) <10th centile were compared with 20 preterm AGA infants. The total duration of respiratory support was recorded. Along with standard ECHO views, the abdominal aorta was imaged.
Results: The GA and BW of FGR and AGA infants were 29.8 ± 1.3 vs. 30 ± 0.9 weeks ($p = 0.78$) and 923.4 ± 168 vs. 1403 ± 237 g ($p < 0.001$). At 10.5 ± 1.3 days after birth, BP was significantly higher in the FGR infants. The aorta intima media thickness (aIMT) was higher in FGR infants (621 ± 76 vs 479 ± 54 μm, $p < 0.001$). The aorta was significantly stiffer in FGR infants. FGR infants also had significantly greater ventricular septal hypertrophy and a lower sphericity index (1.53 ± 0.15 vs 1.88 ± 0.2, $p < 0.001$). Diastolic dysfunction was noted (↑Transmitral E/A ratio, ↑Tei index, and ↑End systolic wall stress [0.84 ± 0.05 vs. 0.78 ± 0.03, $p < 0.001$, 0.34 ± 0.04 vs 0.25 ± 0.03, $p < 0.001$ and 45.9 ± 6.9 vs. 33.7 ± 4.2 g/cm2, $p < 0.001$]). The mean velocity of circumferential fibre shortening (mVCFc) was reduced in FGR infants (1.93 ± 0.4 vs 2.77 ± 0.5 circ/s, $p < 0.001$). Total duration of respiratory support was significantly longer in FGR infants and correlated with ECHO parameters; E/E' ($r = 0.65$, $p = 0.001$) and mVCFc ($r = -0.64$, $p = 0.001$). Significant correlations between vascular biomechanics and cardiac function were noted (Resistance vs E/E' = 0.7 & Tei index = 0.79).
Conclusions: Preterm FGR infants have impaired cardiac function which is associated with respiratory sequelae.

P1157 - CONGENITAL HEART DISEASE DOES NOT INCREASE THE RISK OF DEATH IN LOW BIRTH WEIGHT INFANTS

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Congenital heart disease (CHD) is recognized as a risk factor for poor outcome in low birth weight infants, but its characteristics have not been fully understood.
Methods: To characterize the incidence and outcomes of CHD in low birth weight infants, we retrospectively reviewed the clinical data of infants weighing less than 2,500 g, who were born and admitted in our NICU between 2010 and 2014. We compared the data between infants with CHD and those without CHD (non-CHD).
Results: We excluded 21 infants with PDA alone, and analyzed the data for 1,825 infants. Of these, we diagnosed 86 infants as having CHDs (4.7%) and identified 52 infants with VSD (60%), 14 with TOF (16%), and 12 with ASD (14%). One infant each with HLHS, TGA, TA, TAPVR, and truncus arteriosus was also included. Chromosomal abnormalities were found in 32 infants (1.8%), and the incidence in the CHD group (30%) was significantly higher than that in the non-CHD group (0.3%) ($P < 0.0001$). Six infants in the CHD group (7.0%) and 45 infants in the non-CHD group (2.7%) died during their hospital stay. The CHD group showed a significantly higher mortality rate than the non-CHD group ($P = 0.047$), and infants with chromosomal abnormality also showed a significantly higher mortality rate than those without chromosomal abnormality (19.4% vs. 2.5%, $P < 0.0001$). A logistic regression analysis revealed that CHD alone was not a risk factor for death (odds ratio [OR]: 0.80; 95% confidence interval [95% CI]: 0.20-3.21) but chromosomal abnormality introduced a significantly high risk (OR: 28.0; 95% CI: 6.2-127, $P < 0.0001$).
Conclusion: Mortality in low birth weight infants with CHD was higher than in those without CHD. This difference was due to the higher incidence of chromosomal abnormalities and not due to the presence of CHD alone.

P1181 - EFFECTS OF PERCEIVED KNOWLEDGE GAIN FROM A NURSE RESIDENCY PROGRAM IN SOUTHERN INDIA

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Background/Hypothesis: There was a gap identified between the nurses' training and clinical expectations in a Pediatric Cardiac Intensive Care Unit (PCICU) in Bangalore, India. To address the identified gap, interventions were implemented through a nurse residency program (NRP) with the aim to enhance nurses' knowledge and patients' outcomes following cardiac surgery. Our study examined the feelings and perceptions of nurses' transitioning into clinical educator roles.
Materials and Methods: The five clinical educators participating in the NRP were chosen via purposeful sampling to participate in focus groups. Structured interviews were recorded and transcribed, and later analyzed by two researchers. One of the researchers was a disinterested member to remove bias from the analysis. Thematic analysis, a rigorous process of analyzing data, establishing codes, and identifying themes, was used to analyze content (Braun & Clarke, 2006). The researchers compared interpretations and common patterns to generate conclusions.
Results: Analysis showed common themes in the clinical educators' perceptions regarding their experience with the NRP. We found the clinical educators perceived the NRP led to an overall knowledge gain among participants, leading to increased confidence level, clinical competence, ability to use voice, ability to guide and teach others, happiness, and clinical care improvement. In addition, the clinical educators felt the knowledge enhancement led to greater recognition and respect from others. We found that these results ran parallel to previous publications and current literature.
Conclusions: Clinical educators felt the NRP had a positive impact on nursing practice and relationships in the PCICU, and the continuation of the program would be beneficial to their future careers.

P1182 - PEDIATRIC CARDIAC INTENSIVE CARE NURSE RESIDENCY PROGRAM'S EFFECT ON NURSING PERFORMANCE BEHAVIORS IN SOUTHERN INDIA

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Background/Hypothesis: We identified a gap between nurses' academic training and Pediatric Cardiac Intensive Care Unit (PCICU) nursing care requirements. A nurse residency program (NRP) was created to improve nursing performance behaviors (i.e., knowledge, skills, competence). The aim of this study was to evaluate the NRP's effect on nursing performance behaviors.
Materials and Methods: The NRP consisted of three one-week training sessions and structured mentoring by on-site clinical educators. Our study was a pre-post cohort investigation which explored the NRP's effects on: 1) knowledge, 2) the mentors' and the NRP participants' perception of their nursing skill, and 3) the PCICU medical director perception of participants' competence. 40 PCICU nurses participated in this investigation (NRP $n = 20$; Controls $n = 20$). NRP participants and controls were asked to complete knowledge tests and skill assessments prior to the initiation of the NRP (T1), upon completion of the NRP (T2), and 6 months after completion of the NRP (T3). The mentors evaluated NRP participants and controls' skill at T1, T2, and T3. The medical director evaluated competency of all participants at T1 and T3. A mixed effect model with repeated measures was used to study differences between cohorts.
Results: There were no significant differences between NRP participants and controls' age, gender, birth state, and professional qualification. Mean NRP knowledge scores were higher than controls at T2 ($p < 0.0001$) and at T3 ($p = 0.01$). The NRP cohort mean knowledge scores were higher at T2 ($p < 0.0001$) and T3 ($p = 0.002$) than T1. NRP participants' perception of skill level were higher than controls at T2 ($p = 0.011$) and T3 ($p < 0.0001$). Mentors' perception of NRP participants' skill were higher

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than controls at T2 ($p < 0.0001$) and T3 ($p < 0.0001$). Physician perception of the NRP participants' clinical competency was higher than controls at T3 ($p < 0.0001$).
Conclusions: There were sustained improvement in nursing performance behaviors for participants in the NRP cohort.

Table 1. Outcomes by intervention group and time.

	T1 Mean (SD)	T2 Mean (SD)	T3 Mean (SD)
Mean knowledge scores (0-100%)	60.82 (10.33)	78.32 (5.71)	72.70 (11.26)
Self-rated skill level (1-novice to 4-trainer)	1.93 (0.53)	2.78 (0.73)	3.04 (0.57)
Mentor-rated skill level (1-novice to 4-trainer)	1.69 (0.38)	2.81 (0.53)	2.72 (0.42)
Physician-rated competency (1-strongly negative to 7-strongly positive)	5.92 (0.34)		6.42 (0.49)

Table 2. Outcomes by control group and time.

	T1 Mean (SD)	T2 Mean (SD)	T3 Mean (SD)
Mean knowledge scores (0 to 100%)	57.55 (9.48)	57.95 (7.25)	61.20 (12.20)
Self-rated skill level (1-novice to 4-trainer)	2.09 (0.74)	2.15 (0.75)	1.78 (0.63)
Mentor-rated skill level (1-novice to 4-trainer)	1.52 (0.25)	1.84 (0.66)	1.78 (0.63)
Physician-rated competency (1-strongly negative to 7-strongly positive)	5.30 (0.59)		5.29 (0.81)

P1183 - OUTCOMES ASSOCIATED WITH IMPLEMENTATION OF A PEDIATRIC CARDIAC INTENSIVE CARE NURSES RESIDENCY PROGRAM IN SOUTHERN INDIA

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Background/Hypothesis: We identified a gap between nurses' academic training and Pediatric Cardiac Intensive Care Unit (PCICU) nursing care requirements. A hospital-based nurse residency program (NRP) was created to improve nursing care delivery to children with structural heart disease in a Southern Indian PCICU. Our study examined NRP participants' satisfaction with the NRP and organizational outcomes associated with implementation of a NRP.
Materials and Methods: An international focus group was formed to create the NRP. The NRP consisted of three one-week training sessions and structured mentoring by on-site clinical educators. Twenty PCICU nurses participated in the NRP. Along with education on nursing care for children with structural heart disease each participant was educated on role-modeling adherence with hospital acquired infection reduction bundles and best ways to support bundle adherence in the PCICU. Nursing turnover rates, surgical volume, mortality rates, central-line associated bloodstream infections (CLASBI), ventilator associated pneumonia (VAP), catheter-associated urinary tract infections (CAUTI), and surgical site infection (SSI) rates were compared seven months prior to implementation of the NRP and seven months after initial NRP implementation. Descriptive statistics and Student t-test were used to evaluate outcome changes after the implementation of a NRP in a resource-constrained environment.
Results: Upon completion of the NRP, participants' mean NRP satisfaction score was 1.4 (SD = 0.41) on a Likert Scale ranging from strongly positive (1) to strongly negative (4). There were positive trends in a number of organizational and child outcomes during and after NRP implementation. The most significant was a reduction in mortality rate ($p = 0.007$). In addition, the PCICU CLASBSI rate was marginally lower after implementation of the NRP ($p = 0.056$).
Conclusion: Participants were satisfied with the NRP and there may have been positive effects on organizational and child outcomes. Further study is warranted.

Table 1. Organization and Child outcomes.

Outcome	Pre-education 7 months Mean (SD)	During and After education 15 months Mean (SD)	t, p
Mortality Rate	4.29 (.58)	3.39 (0.70)	t(20) = 3.14, 0.007*
CLASBSI Rate	6.09 (3.42)	3.0 (1.65)	t(20) = 2.26, 0.056*
Nursing Turnove Rate	11.86 (6.31)	6.93 (3.99)	t(20) = 1.89, 0.093
Pediatric Surgical Volume	203.14 (15.92)	223.67 (9.81)	t(20) = 1.60, 0.12
VAP Rate	2.6 (5.6)	0 (2.5)	t(20) = 1.67, 0.14
CAUTI Rate	0 (0)	0.1 (0.38)	t(20) = -1.0, 0.33
SSI Rate	1.35 (0.99)	2.15 (1.52)	t(20) = -1.47, 0.16

P1184 - DEVELOPMENT AND IMPLEMENTATION OF A PEDIATRIC CARDIAC CRITICAL CARE NURSE RESIDENCY PROGRAM IN BANGALORE INDIA

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Background/Hypothesis: Many nurses in resource constrained environments do not receive subspecialty pediatric cardiac

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intensive care (PCICU) training. Developing a sustainable, reproducible Nurse Residency Program (NRP) may address the gap between academic preparation and clinical practice in the Pediatric Cardiac Intensive Care Unit (PCICU) and improve nursing care of children requiring cardiac surgery.
Materials and Methods: A international focus group of pediatric cardiac nursing experts, in collaboration with Children's HeartLink, developed three peer-reviewed one-week PCICU nurse training sessions. The sessions focused on nursing management of post-operative pediatric cardiac surgical patients, implementation of hospital acquired infection reduction bundles, and methods to foster improved communication skills amongst healthcare team members. The NRP contained didactic, low-fidelity simulation, and mentored clinical care training components. NRP presentations encompassed a variety of topics included but limited to cardiac anatomy and physiology, review of critical organ systems, electrophysiology, and congenital heart defect specific nursing care. In addition, we developed a peer-reviewed structured mentor training program for expert local nursing staff. The NRP was implemented in an 81 bed PCICU in Bangalore, India during May-July 2015. Mentors received in depth coaching for our focus group weekly in order to prepare the mentors for future independent implementation of the NRP at our local site.
Results: Twenty-two nurses participated in the initial NRP. Six nurses were trained to become mentors and/or NRP coordinators. Subsequently, a second cohort was successfully led by the mentors January-March 2016.
Conclusions: A NPR can be implemented in resource constrained environments by local teams with the support a nongovernmental organization and expert nurse volunteers. A sustainable, reproducible model of PCICU may be a viable option for improving nursing care.

P1185 - UTILIZATION OF A REDCAP DATABASE AS A NOVEL TEACHING TOOL IN NURSE RESIDENCY PROGRAM

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Background/Hypothesis: We discovered an educational gap between nurses' academic training and their clinical practice requirements for caring for critically ill children with congenital heart defects. Cincinnati Children's Hospital Medical Center, Children's HeartLink, and Narayana Institute of Cardiac Sciences (NICS) in Bangalore, India developed an intervention to improve Indian child outcomes after cardiac surgery through development of nursing expertise in a pediatric cardiac intensive care unit (PCICU).
Materials and Methods: Our team addressed this gap through development and implementation of a hospital-based PCICU nurse residency program. In addition, Children's HeartLink clinicians developed a comprehensive 75 question cardiac nursing examination in a REDCap database to assess the PCICU nurses' knowledge pre- and post-residency program. The multiple choice examination was emailed to each nurse participant utilizing the survey function in REDCap and was scored for commonly missed responses. Subsequently, respondents and mentors were notified of areas for further teaching.
Results: The REDCap database has proven to be an excellent teaching tool with the ability to improve nursing expertise. Median testing scores increased from 62.66% to 77.33%, showing an overall increase in scores of greater than 23%. The REDCap database provided a robust dataset and offered immediate feedback for the investigator that allowed for real-time assessment of the participating nurse's learning and adjustment of teaching strategies in order to promote participant learning.
Conclusions: The REDCap database has proven to be a novel tool in fostering nursing expertise that in turn may positively affect child outcomes at NICS.

P1226 - QUALITY IMPROVEMENT MINIMIZING UNPLANNED EXTUBATION IN PEDIATRIC INTENSIVE CARE UNIT

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Background: Unplanned extubation (UE) is a removal of the endotracheal tube by action of patient (self- extubation) or dislodge during care and manipulation of the patient. UE can cause serious complications for mechanical ventilation patient and it is one of the indicator in determining the quality of care in Pediatric Congenital Heart Centre, Intensive Care Unit (PCHC ICU) of National Heart Institute, Malaysia.
Objectives: To identify the risk factors of the incidence with the unplanned extubation and to evaluate the effectiveness of the targeted interventions with a reduced rate of the unplanned extubation.
Materials and Methods: Clinical information, incidence reports and demographic data of the patients were collected retrospectively over a 3-year period from January 2014 until October 2016 related to the unplanned extubations. Key performance indicator, continuous nursing educational programs, new securing techniques of ETT and protocols on care management were intervened and implemented to reduce the rate of unplanned extubations. Results: In year 2014 total number of 31 incident has occurred. After implementations of few targeted intervention, there were reduction to 10 incidences in the year 2015. From January 2016 till October 2016 the number of events was 11. Out of total incident for past 2 years till October 2016, the major factors that identified were due to patient factor about 51.6%.
Conclusion: Risk factors of unplanned extubation are varies. Based on identified risk factors, it contributes in implementing preventive measures to ensure the reduction of events in promoting patients safety. Good practice and the targeted interventions support in reduction of these potentially adverse events.

P1235 - DIRECT ECHOCARDIOGRAPHIC IMAGING OF THE BERLIN HEART VALVES CAN AID IN DIAGNOSING VALVE DYSFUNCTION

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Introduction: Berlin Excor is a VAD that has been approved by the FDA for use in pediatric patients. The Berlin Heart (BH) differs

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from many mechanical circulatory support (MCS) devices in that it is a pulsatile pump with two sets of artificial valves within the pumping mechanism. The primary drawback of the BH is a 20-30% stroke risk. This high rate comes in large part from thrombus formation on the valves. The BH valves are polyurethane (PU) tri-leaflet valves that significantly reduce backflow compared to mechanical valves. We report a novel way of directly imaging the BH valves that can lead to early thrombus detection and potentially decrease the risk of stroke. Imaging for valve regurgitation can also help detect loading conditions.
Method: Accurate images of inflow and outflow valves can be obtained bedside using a standard echocardiogram machine. Ultrasound gel is applied to the transducer and the transducer is placed directly onto the BH valve casing. The valve leaflets can be seen in 2D imaging and with color flow. Thrombi are difficult to visualize by 2D imaging alone but changes in color flow pattern representing increased valve regurgitation from thrombus formation can easily be seen.
Discussion: BH remains one of few options for infants and young children who require MCS. Unfortunately it carries a high risk of ischemic stroke. We present a new technique to monitor for the development of thrombus before it is visible. We have utilized this technique on several patients and believe there is an association between increasing valve regurgitation and need for pump exchange secondary to thrombus formation. Echocardiography can detect changes in valve regurgitation which should increase suspicion for early thrombus formation, which could lead to a decrease incidence in strokes. Direct imaging of BH valves can also detect changes in the afterload leading to regurgitation.

P1257 - CARDIAC RECOVERY AND OUTCOME OF NEONATES AND INFANTS PRESENTING WITH SEVERE AORTIC COARCTATION AND DEPRESSED CARDIAC FUNCTION

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Coarctation of the aorta (CoA) accounts for 5-8% of all congenital heart defects. Children with severe CoA may present with significant myocardial depression and shock. The aim of this study to identify short and midterm outcome of neonates and infants diagnosed with isolated Coarctation and depressed cardiac function (CoA-DF) looking for their cardiac recovery and overall outcome after repair.
Methods: All cases of isolated coarctation who had surgical repair between December 2002 and December 2015 were reviewed retrospectively. A case control study was performed comparing CoA-DF cases defined as shortening fraction (SF) < 25%) and ejection Fraction (EF) < 55% with cases of CoA and normal cardiac function (CoA-DF V.S control groups). Indices of cardiac function before and after repair were reviewed and statistically compared as well as outcome in both groups.
Results: 58 cases fulfilled inclusion criteria with 25 subjects in CoA-DF group and 33 in control group. There were statistically significant difference in EF (42 ± 14.6 v.s 66 ± 8.7) and SF (20 ± 7.6, 36 ± 6.7) pre surgery between both CoA-DF and control groups respectively ($P < 0.05$). Cardiac indices and ventricular function showed improvement in cases of CoA-DF within six months post repair to become comparable to control group. There was no difference in term of complication between both groups.
Conclusion: Majority of the patients with isolated CoA-DF showed improvement of function within 4 weeks after surgery. Patients with residual CoA or complications such as pulmonary hypertension (PHT) has longer recovery extending up to 6 month after surgery. Delayed recovery may hint to the presence of residual lesion or complications that may affect the myocardial ability to recover normally.

P1263 - MANAGEMENT OF CHYLOTHORAX IN CHILDREN AFTER CONGENITAL HEART SURGERY

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Background: Chylothorax is one of the serious complications after cardiothoracic surgery in children and usually manifests during the first postoperative week. Its appearance prolongs hospital stay and increases morbidity. A large number of endogenous proteins are lost, including those involved in the coagulation cascade, increasing the risk of venous thrombosis, malnutrition, infections and other complications.
Method: The study included 19 patients, with a median age of 2,7 months (SD +-1,6) between April 2013 and May 2015. All patients were treated according to our local protocol. The responses of patients to nutritional modifications, octreotide therapy, anticoagulant management and surgical interventions were reviewed.
Results: The prevalence of postoperative chylothorax, which developed at a median of 8 days after operation (3 to 26 days), was 6,1%. Surgical procedures included: repair of atrioventricular septal defects in 5 (26%), Jatene surgery in 4 (21%), single ventricle staging in 5, correction of Fallot in 3, double switch 1 and correction of total anomalous pulmonary venous drainage 1. The median duration and total volume of chylous drainage was 15 days (5 to 26 days) and 6,4 to 68 mL/kg/day. All patients received heparin prophylaxis and only one developed venous thrombosis. 16 patients responded to conservative therapy (84%). Octreotide was used in 14 patients (73%). Only three (15%) had to undergo surgery (thoracic duct ligation and pleurodesis). No patient died due to chylothorax. Only one patient developed venous thrombosis.
Conclusions: Conservative therapy has proven to be effective in most cases, reserving surgical treatment only for refractory cases. The use of octreotide decreases days of pleural effusion in half (13 vs 27 days), if it is used early (before 15 days of surgery), and Heparin prophylaxis proved to be effective in reducing the rate of associated thrombosis.

P1272 - PACKED RED BLOOD CELLS DILUTED WITH 0.9% NACL INFLUENCE ON OSMOLALITY TOTAL HEMOGLOBIN AND MARKERS OF HEMOLYSIS LEVELS

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Background: Dilution of packed red blood cells (RBC) with 0.9% NaCl is performed in order to reduce blood viscosity and increase infusion rate. Despite contraindications, nurses and physicians analyze the risks and benefits in critical situations before deciding to manipulate the RBC, but the effects of this procedure on RBC

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characteristics is indefinite. The objective of this study was to describe the osmolality of RBC diluted in 0.9% NaCl and to verify the effects on total hemoglobin, hematocrit, free hemoglobin, and hemolysis levels.
Materials and Methods: An experimental study approved by the ethical committee was developed with nine packed RBC (08 A+ and 01 AB+), storage time of 17 ± 8.9 days. The proportions of 0.9% NaCl dilution studied were 25% and 50%. Samples were collected directly of the packed RBC and after dilutions. Osmolality(mOsm/Kg) was measure by an osmometer; total hemoglobin(g/dl) and free hemoglobin (g/dl) were determined through spectrophotometry, hematocrit (%) and hemolysis degree (%) (free hemoglobin × 100-hematocrit/total hemoglobin) were calculated. Data were analyzed according to mean ± SD and ANOVA test ($p \leq 0.05$).
Results: Osmolality in the packed RBC was 350 ± 7.95; 336 ± 7.87 after dilution at 25% and 313 ± 3.52 at 50% ($p < 0.0001$). The initial level of total hemoglobin was 30.05 ± 5.59, with 25% 0.9% NaCl dilution decreased to 22.37 ± 4.93 and 14.06 ± 3.33 at 50% ($p < 0.0001$). Hematocrit values ranged from 73.6 ± 3.26, to 59.5 ± 3.23 at 25% dilution and 39 ± 2.35 at 50% ($p < 0.0001$). The levels of free hemoglobin was 0.06 ± 0.03; 0.11 ± 0.16 after dilution at 25% and 0.10 ± 0.15 at 50% ($p = 0.7025$). The initial degree of hemolysis was 0.06 ± 0.04; 0.25 ± 0.48 after dilution at 25% and 0.46 ± 0.66 ($p = 0.2324$).
Conclusion: There was a significant decrease on the osmolality, total hemoglobin and hematocrit levels, as expected. Hemolysis markers showed increased levels with no statistically significant variations.
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P1276 - PULSE WAVE TRANSIT TIME USED IN THE PERIOPERATIVE PERIOD OF CONGENITAL HEART DISEASE

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Back ground: The aim of this study is to assess whether pulse wave transit time (PWTT) is able to assess stroke volume in the children with congenital heart disease.
Method: This is a retrospective study of 56 consecutive patients with congenital heart disease who underwent definitive repair between July 2013 and March 2016. Exclusion criterions are palliative procedure and sustained arrhythmia. Pulse wave transit time and echocardiography were measured at perioperative day, postoperative day 1, and day 7. PWTT was measured and corrected data by body surface area was defined as PWTT index. Stroke volume was measured in the short axis view by transthoracic echocardiography. Patients were divided into two groups (group A: age ≧ 1 year, group B: age < 1 year).
Results: The median age and body surface area at surgery was 12 months (range; 13 days - 16years) and 0.44 m2 (minimum; 0.16 m^2 -1.52 m^2). The correction coefficient between PWTT index and stroke volume was $r = -0.427$ ($p < 0.01$) in a total of 140 data sets for 56 cases. In group A, the correction coefficient was $r = -0.488$ ($p < 0.01$) in a total of 95 data sets for 32 cases. Whereas, the correction coefficient was $r = -0.065$ ($p = 0.67$) in a total of 45 data sets for 24 cases in group B.
Conclusions: PWTT index as a non-invasive parameter was associated with stroke volume with children in the perioperative period of congenital heart disease. Although, it was still not be enough for the the parameter in neonates and infants.

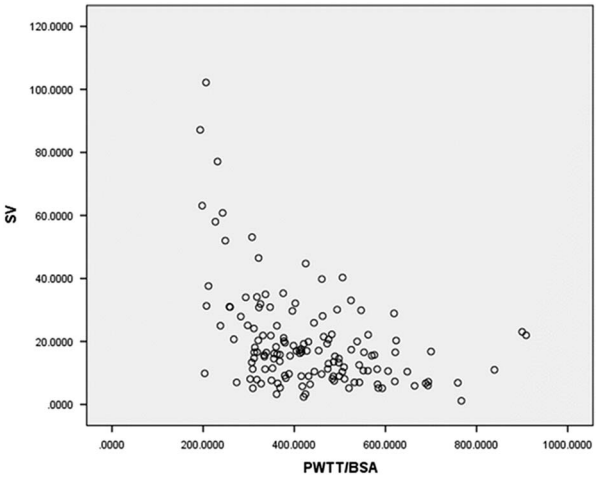

Figure.

P1283 - REDUCING OPIOID USE IN THE PEDIATRIC CARDIAC INTENSIVE CARE UNIT A QUALITY IMPROVEMENT PROJECT

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Background: Sedation and analgesia reduce distress, ease mechanical ventilation, reduce metabolic needs, and facilitate stable hemodynamics. Excessive sedation can lead to longer stays, significant morbidities/mortality, withdrawal syndromes, and long-term neurological/behavioral difficulties. Protocolized sedation for ventilated adult patients has been shown to improve outcomes like morbidity, length of stay, and duration of ventilation, but pediatric data is limited. We designed and implemented a standardized comfort algorithm for a 25-bed CICU at Cincinnati Children's Hospital to ensure adequate comfort while minimizing use of opioids.
Materials and Methods: We adapted a PICU sedation protocol (Deeter et al., 2011) for children after elective cardiac surgery. We selected dexmedetomidine as a first-line sedative/anxiolytic agent rather than midazolam. We employed a dedicated opioid infusion line (separate from vasoactives) to facilitate delivery of as-needed morphine bolus doses from the infusion pump, obviating the need for the bedside nurse to leave the bedside of a distressed child to obtain medications and find a second nurse for double checking. A multidisciplinary team employed the Model for Improvement to refine the algorithm over nine months based upon detailed review of successes and failures, local cultural concerns, practice-related barriers and multidisciplinary feedback. Process, balancing and outcome measures were studied using statistical process control methodology.
Results: Over 12 months following implementation, the morphine infusion rate for all ventilated children was reduced from a baseline

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monthly rate of 120 to 51 mcg/kg/hour (58% overall reduction). Unplanned tracheal extubation, duration of mechanical ventilation and length of CICU stay were unaffected.
Conclusions: Use of a comfort algorithm reduced infusion doses of opioid medications in our CICU. Duration of mechanical ventilation and length of ICU stay did not change, which may reflect CICU cultural factors and stepdown unit bed availability. Reducing opioid usage did not affect safety as judged by a stable rate of unplanned extubation.

P1286 - UNUSUAL CASE OF INTERVENTRICULAR SEPTAL HAEMATOMA AFTER SURGICAL REPAIR OF FALLOT TYPE DOUBLE OUTLET RIGHT VENTRICLE

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Background: Interventricular septal haematoma (IVSH) is a very rare complication after congenital heart surgery in childhood. Since 2005 there are 11 cases reported in the literature. In all of these cases with two exceptions, the surgical correction includes ventricular septal defect (VSD) closure and IVSH probably results from damage of the perforating branches of the coronary arteries near the suture lines of the VSD patch. IVSH manifestations may vary from mild heart failure to low cardiac output states (LCOS) and disturbances of the heart rhythm. The diagnosis is set by an echocardiography. There is no single treatment strategy. It is considered that secondary obstruction of ventricular inflow or outflow and severe systolic dysfunction are indicators for surgical treatment.
Aim: To present an unusual case of IVSH after congenital heart surgery, in which the IVSH is not related to the VSD patch closure.
Methods: Retrospective analysis of the medical files of the patient.
Results: We present a 6 months old boy after surgical repair of Fallot-type DORV with augemtation of right ventricular outflow tract (RVOT) with transannular patch (TAP). Early postoperative period was complicated by LCOS and third-degree AV block. Transesophageal echocardiography showed a thickening of the ventricular septum, moderate depression of the LV systolic function, no obstruction of both ventricular inflows and outflows. No surgical treatment was considered. Patient died at the end of first week with severe respiratory distress syndrome (RDS) on ventilator. Autopsy data showed RDS associated with respiratory syncytial virus (RSV) infection, large 10/12mm fusiform IVSH, originating near the proximal sutures of the transannular patch and not connected to the VSD patch.
Conclusion: IVSH is a rare complication after congenital heart surgery and in most cases is associated with VSD closure, but could be a result of TAP augmentation of RVOT.

P1298 - POSTOPERATIVE COMPLICATIONS IN PEDIATRIC CARDIAC SURGERY PROGRAM WITH LIMITED SYSTEMIC RESOURCES

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Background: The complication rate after pediatric heart surgery, as well as the failure-to-rescue rate are less known in countries with limited systemic resources.
Methods: The postoperative complication (POC) rate, mortality related to complications, 25 patient and procedure variables as risk factors for developing POC or death and 27 POCs as predictors of mortality were evaluated.
Results: This retrospective, single-center observational study constituted 296 consecutive patients under 18 years of age, who underwent surgical interventions for congenital heart disease in Kharkiv cardiac center (Ukraine) between January 2014 and December 2015. Overall mortality was 5.7%. There were no deaths among 223 patients without POCs. The remaining 73 patients (24.7%) developed POCs and had 17 mortalities (23.3%). There was a strong association between number of POCs and mortality - from 8.1% among patients with only one POC to 35.3% with two POC to 42.1% with three and more POCs ($p = 0.007$). Risk factors for developing POC were younger age, previous cardiac surgery, extracardiac structural anomalies, concomitant diseases and cardio-pulmonary bypass, whereas total number of complications per patients was the only predictor of death. Among POCs a postoperative low cardiac output and acute renal failure requiring replacement therapy were independent predictors of mortality.
Conclusions: Rate of POCs after pediatric cardiac surgery in program with limited resources was less than in high-income countries, due to less complex patients. Despite the comparable crude mortality, the mortality among patients with POC (failure-to-rescue) in our series was significantly higher than in developed world. A number of initiatives are needed in order to improve failure-to-rescue rates after pediatric cardiac surgery in developing countries.

P1306 - VASOACTIVE INOTROPIC SCORE AND OUTCOME ASSESSMENT IN CYANOTIC INFANTS AFTER CARDIOVASCULAR SURGERY

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Background: Infantile cardiac surgery has made significant progress in last few decades but to compare various surgical modalities and treatment protocols, we lack one objective prognosticating criteria. This prospective clinical assessment of Vasoactive inotropic score (VIS) was undertaken to predict post operative outcomes in cyanotic infants after cardiopulmonary bypass surgery.
Methods: Post operative outcomes were analyzed in 100 cyanotic infants (less than 1 yr age) operated between September 2014 to April 2016. Inotropic score (IS) and Vasoactive inotropic score (VIS) were calculated for all patients at 1 hr, 24 hr and 48 hr after surgery.
Results: Mean surgical age was 7.39 months (3 days to 12 months) with 85% infants being male (85/100). In 81% patients intracardiac repair was performed (TAPVC 40, TOF 24, DORV VSD PS 4 and others 13) while rest 19% patients had single ventricular physiology requiring bidirectional Glenn operation. On regression analysis, prolonged ICU and hospital stay correlated best with VIS score at 48 hr interval ($R2 = 0.67$ and 0.50 respectively). Low birth weight (<2.5 kg), requirement of pre operative ventilation and high 1st hr VIS score (>25) were found predictive of higher post operative mortality. Maximum VIS score at 24 hr and 48 hr, on Spearman analysis were shown positively correlated with adverse post operative outcome.

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Conclusion: In cyanotic infants undergoing cardiac surgery, higher VIS score at 48 hrs was strongly associated with poor post operative outcome and is a good assessment tool for requirement of resources in post operative care.

P1317 - ARTERIAL UMBILICAL CATHETER FRAGMENT EMBOLIZATION IN AN INFANT

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Background: Catheterization of the umbilical vessel is a standard procedure in neonatal ICU, especially in premature infants. Rarely, the umbilical arterial catheter (UAC) may break leaving fragments that can provoke thrombosis, vasospasm, infection, distal embolization and even death.
Methods: Premature male infant, 24 weeks, 800 g, remained hospitalized for three months during which he was submitted to surgical procedures for the ductus arteriosus closure and necrotizing enterocolitis. Readmitted three days after discharge with hypoactivity and apnea, suggestive of sepsis, progressed with temperature decrease and distal cyanosis of left lower extremity (LLE). Doppler ultrasound confirmed emboli of left internal iliac artery. A 6-hour/day intravenous infusion of rTPA (0,2 mg/kg/h) was performed but suspended on day four due to nosebleed. Abdominal ultrasound detected catheter fragments in the iliac artery, which were removed by a transumbilical approach. Due to progressive distal necrosis of LLE 15 sessions of hyperbaric oxygen therapy (HBOT) were implemented, and enoxaparin (1,5 mg/kg/dose 12/12 h) was administered for a month, then substituted by acetylsalicylic acid (5 mg/kg/day). Later, the patient was submitted to debriding of LLE toes' necrotic tissue, and a heel culture was positive for Pseudomonas.
Results: Despite all efforts, resection of the LLE toes up to metatarsus was necessary, but below-knee amputation, initially recommended, was avoided. The preservation of the heel allowed physical rehabilitation and future prosthetic use.
Conclusion: Although UAC transection is rare, it can be lethal. Even after removal of the fragments the risk of limb ischemia remains; also, thrombolytics and anticoagulants can cause intracranial hemorrhaging. The rTPA and enoxaparin associated with HBOT proved to be efficient in this case. It is recommended that the removed catheter be always measured to avoid, in the event of a fracture, the serious complications caused by remaining fragments in the child's body.

P1336 - FUNCTIONAL ECHOCARDIOGRAMS IN PEDIATRIC INTENSIVE CARE UNIT

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Introduction: Functional Echocardiography has shown to impact medical decision by detecting cardiac dysfunction early in recent adult studies. The purpose of this study was to evaluate the cardiac functions by bedside 2D Echocardiogram/ Color Doppler in critically ill children in pediatric Intensive care Unit (PICU).
Methods: It is a descriptive, observational, prospective case control study in a tertiary care Children's Hospital. 83/100 critically ill PICU children who fulfilled the study criteria were included. Initial Echocardiogram was done within 48 hrs of admission and repeat echocardiograms if necessary. Cardiac dimensions, functions and cardiac output were assessed by standard methods and compared to pediatric norms for age and Body Surface area (BSA). Volume status was assessed by inferior venacaval (IVC) collapsing index (IVC-CI) and IVC distensibility index (IVC-DI).
Results: Left ventricular hypertrophy (LVH) was noted in 22.9% and high LVMI was noted in children with abnormal renal profile, hypertension and hypocalcaemia [$p < 0.05$]. LV systolic dysfunction was significant in sepsis group patients (32.5%) [$p = 0.0063$] and those who had a PICU stay more than 10 days duration. [$p = 0.013$]. Isolated LV diastolic dysfunction was found in 88.7% with (61.9%), having preserved LV systolic function [$p = 0.0058$]. Abnormal RV function was noted in (68.7%). Low LVCO-index was noted in (45.0%) and high in (21.7%). The IVC-CI more than 55% was found in (26.5%) and IVC-DI more than 18% (91.6%) suggestive of fluid responsive states. During PICU course ,after a 2D Echocardiography evaluation 69.9% patients ($n = 58/83$) required medical interventions. Ten (12%) children expired during study period.
Conclusion: Evaluation of Cardiac functions and volume status by functional echocardiography helps to tailor fluids and inotropes and support hemodynamics in critically ill children in PICU.

P1345 - THE ASSESSMENT OF THE ORAL FEEDING BEHAVIORAL INTERVENTION PROTOCOL IN THE NEONATES AFTER ARTERIAL SWITCH PROCEDURE

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Objective: To improve newborns' feeding ability and maternal feeding skills after arterial switch procedure through the feeding behavioral intervention protocol.
Methods: A before-after study was conducted on 140 newborns and their mothers who were divided into experimental group and control group. Outcomes including newborns' feeding behavior and mothers' satisfaction, self-efficacy and depression were be Compared and analyzed.
Results The oral motor score was improved ($p < 0.05$) and the length of gastric usage was shortened ($P < 0.01$) in the experimental group; the maternal satisfaction score, self-efficacy score and the depression score was improved ($p < 0.01$)in the experimental group compared to those in the control group.
Conclusion: The reasonable and scientific intervention protocol improves patients' feeding ability and the quality of life after arterial switch procedure and it also improves maternal feeding skill and comfort their emotion.

P1357 - ENHANCED HANDOFF PROCESS IMPROVES COMMUNICATION LEADING TO IMPROVED PATIENT SAFETY AND OUTCOMES

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Background: Congenital heart surgery patients, who can deteriorate quickly, moving from the operating room to the intensive care unit requires co-ordination of people, devices, information, and patient knowledge, under time pressure,. We employed a range fo strategies (task design, task allocation, task sequence, teamwork, communication, checklists, protocols, environment and workspace considerations) to improve teamwork, reduce risk and improve safety.
Hypothesis: A theoretically robust handoff process would create a faster, smoother process with reduced subsequent treatment problems
Patients and Methods: Two distinct phases of care, GROUP 1 were cared for prior to starting the CHPICU prior to establishing the handoff process, and GROUP 2 were cared for in the CHPICU after robust handoff process was developed. 29 patients were included in the study, GROUP 1 n = 16, and GROUP 2 n = 13. Through direct observation, flow disruptions in 5 categories (organization, team, patient, equipment, communication), and clincal care concerns after 6 hours in 5 categories (vitals, bleeding, general clinical, respiratory, and drugs), were evaluated. Results: A proceduralized team handoff was used in 21 patients (72%), 8 did not follow a recognised procedure.Time (min) to complete the handoff was significantly less in Gp 2 (mean = 23.5 ± 7.5, median = 20), compared to Gp 1 (mean = 35 ± 14.6, median = 35), p = 0.029. Flow disruptions (Mean Gp1 = 1.5, mean Gp2 = 0.8, p = 0.19) and treatment problems (Mean Gp1 = 0.9, mean Gp2 = 0.2, p = 0.04) were reduced after implemtation of the process. Vitals and respiratory problems were substantially reduced in Gp 2(36% vs 7%; 21% vs 0%). Equipment problems during the handoff remained a problem in both groups.
Conclusion: A structured handoff process improves detection of missing information, reduces handoff duration, flow disruptions and clinical disruption. Developing a team handoff process will be important in reducing the variablity of care and responding to deterioration and other patient treatment demands.

P1362 - TARGETED PROPHYLACTIC HEPARIN INFUSIONS SIGNIFICANTLY REDUCE CATHETER ASSOCIATED THROMBOSES IN INFANTS AFTER CARDIAC SURGERY

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Background: Infants with central venous catheters (CVC) after cardiac surgery are at an increased risk of venous thrombosis. Previous studies of fixed untargeted heparin infusions (10units/kg/hr) or other anticoagulants have not decreased the incidence of thromboses. We hypothesized that a targeted prophylactic heparin infusion (TPHI) protocol would reduce the incidence of catheter-associated thromboses in infants after cardiac surgery.
Materials and Methods: Beginning in September of 2010, all infants (age less than 6 months or weight less than 5 kg) with an upper extremity CVC received a TPHI for the duration of CVC usage. Heparin infusions were titrated to achieve slightly subtherapeutic unfractionated heparin levels between 0.2-0.3units/mL. All venous ultrasounds obtained between CVC insertion and one week after CVC removal were reviewed for presence of thromboses. Rates of venous thromboses were compared to historical controls receiving untargeted heparin infusions or no anticoagulation.
Results: Of 526 patients (TPHI: n = 397, control: n = 129), 544 CVC were analyzed. The median duration of CVC usage (TPHI = 4.9 days vs. control = 4.0 days, p = 0.06) was similar between cohorts; however, there were fewer venous ultrasounds (22% vs. 33%) in the TPHI cohort. TPHI was associated with a significantly decreased incidence of thromboses per CVC (8.6% vs. 19.1%, $p < 0.01$) as well as a significant reduction in thromboses per 1000 line days (8.0 vs. 19.2). Further analysis of ultrasound data also showed that, among identified thrombotic events, there were fewer occlusive thrombi after implementation of the TPHI protocol (3.5% vs. 12.2%, $p < 0.01$).
Conclusions: Targeted prophylactic heparin infusions significantly decreased catheter-associated thromboses in infants with congenital heart disease.

P1392 - QUALITY IMPROVEMENT INITIATIVE TO REDUCE CENTRAL LINE ASSOCIATED BLOOD STREAM INFECTION IN CARDIAC PEDIATRIC INTENSIVE CARE UNIT

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Background: Central line associated blood stream infections (CLABSI)are among top 10 of death caused by healthcare associated infections. CLABSI resulted in prolonged hospital stays, increased hospital cost, significant morbidity and mortality. The objective of this study is to reduce CLABSI in Pediatric Intensive Care Unit (ICU).
Methods: All patients in pediatric ICU with central line up to 18 years old were included in this study from January to November 2016. CLABSI was defined as per Centers fro Disease Control and Prevention guidelines. Pre intervention rate used as baseline rate.
Interventions were implemented as below: a) Infection Control Link Nurses (ICLN) conducted an observational audit of the nurses on their compliance to the bundle. This allowed ICLN to immediately educate the staff. b) Emphasized on scrub the hub for 15 seconds before give injection was incorporated. c) Empowering nurses to stop the central line insertion procedure if the doctor breaches aseptic technique. d) Introduction of a central line checklist to ensure proper aseptic technique were performed and documented. e) Easy access and availability of a fully equipped central line cart with appropriate consumable needed for central line insertion. f) Increased education by bedside teaching, visual aide at the bedside such as central line bundle poster and line care. g) Daily bath with Chlorohexidine.
Results: CLABSI rates from January to November 2016 was reduced to 1.3/1000 line days compared to 1.7 in 2015 and 5.1 in 2010. We also achieved zero CLABSI in month of May, June, July, October and November.
Conclusions: Multiple interventions were required to reduced CLABSI and it can be achieved with high compliance and commitment. Continuous audit, bedside teaching, education and awareness is very important to ensure sustainability. Interventions such as introducing the checklist, central line cart and empowerment were able to reduce CLABSI in pediatric ICU.

P1403 - PROLONGED MECHANICAL VENTILATION AFTER UNIFOCALIZATION SURGERY IN CHILDREN WITH TETRALOGY OF FALLOT PULMONARY ATRESIA AND MAJOR AORTOPULMONARY COLLATERALS (TOF PA MAPCAS) IS ASSOCIATED WITH NEED FOR FEEDING TUBE AT HOSPITAL DISCHARGE

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Background: Children with Tetralogy of Fallot, Pulmonary Atresia and Major Aortopulmonary Collaterals (TOF/PA/MAPCAs) are at risk for prolonged hospitalization after unifocalization surgery. Feeding problems after congenital heart surgery are associated with longer hospital stay. We sought to determine the impact of baseline and intraoperative factors and duration of mechanical ventilation (VENT) on the need for a feeding tube (FT) at the time of hospital discharge.
Methods: We included children with TOF/PA/MAPCAs undergoing primary or revision unifocalization + /-VSD closure from age 3 months to 4 years from 2010-2016. We excluded patients requiring FT prior to surgery. Children discharged with an enteric tube for supplemental or primary nutrition were included in the FT group. VENT beyond postoperative day 5 was considered prolonged. We compared the FT to non-FT group by uni- and multivariable logistic regression.
Results: Of 56 patients studied, 41% required FT. Median age (7.1 months (Q1-Q3 5.6-16.0) vs 8.8 (6.5-21.5), P = 0.4) and weight z-score (-2 vs -1.5, P = 0.92) were similar in the two groups. A chromosome 22q11 deletion was associated with FT (41% vs 17%, P = 0.05), but surgical type was not (P = 0.26). Right ventricle to left ventricle pressure ratio (RV/LV) was similar in both groups (0.36 vs 0.35, P = 0.33). Median cardiopulmonary bypass (CPB) time in the FT group was longer (335 vs 244 minutes, P = 0.04). Median VENT duration was 4 days (Q1-Q3 2-5.5), and prolonged VENT was associated with FT (48% vs. 9%, P = 0.001). On multivariable analysis, prolonged VENT was the only variable associated with FT (9.2, 95% CI (1.5-56.1)).
Conclusion: Among children with TOF/PA/MAPCAs who were feeding by mouth prior to surgery, prolonged mechanical ventilation after unifocalization surgery was associated with need for FT at discharge. Anticipation of feeding problems in this population and earlier FT placement may help reduce hospital stay.

P1404 - CHILDREN WITH 22Q11 CHROMOSOME DELETION HAVE WORSE EARLY POSTOPERATIVE OUTCOMES AFTER UNIFOCALIZATION SURGERY COMPARED TO THOSE WITH NORMAL GENOTYPE

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Background: A 22q11 chromosome deletion (DEL) is common in children with Tetralogy of Fallot, pulmonary atresia and multiple aortopulmonary collaterals (TOF/PA/MAPCAs). We sought to determine whether patients with TOF/PA/MAPCAs and DEL are at risk for worse early postoperative outcomes including duration of mechanical ventilation (VENT), duration of intensive care unit (ICU) and hospital stay (LOS) following unifocalization surgery.
Methods: We included all patients with TOF/PA/MAPCAs undergoing primary or revision unifocalization + /- ventriculoseptal defect (VSD) closure at our institution from 2008-2016 and excluded patients with unknown DEL. We compared perioperative outcomes using uni- and multivariable regression adjusting for VSD closure and delayed sternal closure (DSC).
Results: Of 180 patients included, 41% had DEL. Need for neonatal surgery (age < 3 months) was similar in children with DEL (P = 0.44). VSD closure was performed concomitantly with unifocalization in 74% of all patients, 67% with DEL (P = 0.06), and cardiopulmonary bypass time (CPB) was similar (P = 0.85). Right to left ventricle pressure ratio (RV/LV) was higher in patients with DEL, 0.37 vs. 0.33 (P = 0.02). DSC was more common in children with DEL, 43% vs. 17% (P = 0.001). Duration of mechanical ventilation was longer in DEL, median of 7 (1-71) vs. 4 (1-40) days (P = 0.001 on unadjusted and adjusted analysis). Duration of ICU stay was also longer in DEL, median of 16 (4-120) vs. 8.5 (2-171) days (P < 0.001 on univariable analysis, P = 0.04 when adjusted for DSC and VSD closure). LOS was not significantly longer in DEL with median of 21 (7-120) vs. 15 (5-174) days (P = 0.07).
Conclusion: Children with TOF/PA/MAPCAs and DEL are at risk for longer duration of mechanical ventilation and ICU stay and higher RV/LV pressures following unifocalization surgery. These data suggest patients with DEL may have less favorable pulmonary arterial anatomy and more postoperative complications.

P1407 - STUDY OF CLINICAL OUTCOMES OF CARDIORESPIRATORY RESUSCITATION IN CHILDREN

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Introduction: Pediatric cardiopulmonary arrest is distinct from adult cardiac arrest. This study aimed at detecting prognostic factors for post CPR outcomes in children.
Methods: Demographic profile, clinical features, laboratory findings, treatment , resuscitation and survival outcome details of 40 children who required in-hospital CPR were analyzed. The outcomes were grouped as long-term survivors (survived to discharge); short-term survivors (survived >24 hours after CPR but not until discharge) and non-survivors (died within 24 hours of CPA). At discharge, patient's neurological functional status was classified as neurologically intact, impaired or dependent.
Results: 82.5% children survived CPR with 68% survivors having normal neurological functional status at discharge. Neither the level of expertise of resuscitator nor the site of CPR affected the outcome. 17(42.5%) required 2 or more CPR efforts during their hospital stay (p = 0.00002). No significant correlation was noted between presenting symptoms or signs to post-CPR survival outcomes (p > 0.05). Those children with oliguria post CPR (25%) and who required volume resuscitation had a poor survival (p = 0.024). Higher motor response post CPR shorter duration of resuscitation, fewer attempts of CPR and early return of spontaneous circulation(ROSC) (87.5%) had a favorable impact for survival (P <0.05). Early enteral feeds within 5 days post CPR in 32(80%) subjects had higher survival rates. (P = 0.0003).
Conclusion: Shorter duration of resuscitation, fewer attempts of CPR, early return of spontaneous circulation, higher motor response and early institution of enteral feeds post CPR had a favorable impact on survival outcomes. Oliguria, increased fluid requirement, hyponatremia, hyperglycemia, hypoxia and hypocarbia in the peri resuscitation period were unfavorable factors for survival. Prolonged ventilator support also correlated with poorer neurological outcome at discharge.

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P1418 - NOVEL STRATEGY FOR PERIOPERATIVE MANAGEMENT OF MODIFIED BLALOCK TAUSSIG SHUNT ON PATENT DUCTUS ARTERIOSUS DEPENDENT CIRCULATION

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Background: Blalock-Taussig shunt (BT shunt) for pulmonary atresia (PA) or severe stenosis (PS) which depends on patent ductus arteriosus (PDA) has still had higher mortality because of post-operative excessive pulmonary flow.
Purpose: Two perioperative strategies for BT shunt of these patients have been employed since 2013. The first strategy was prophylactic decrease or withdrawal of Lipo-prostaglandin E1 (LipoPGE) one week before operation to avoid cardiac failure by reducing preload (low pulmonary flow strategy) and controlled SpO2 level as less than 80%. The other was administration of PDEIII inhibitor from anesthetic induction to control Qp/Qs by reducing afterload (low vascular resistance strategy). The aim of this study was to seek the efficacy of these strategies.
Methods: Forty two patients with PA (n = 34) or PS (n = 8) associated with PDA dependent circulation underwent BT shunt without extracorporeal circulatory support from July 2008 to November 2016. Patients were devided into two groups; No strategy group (N group, n = 20) and Strategy group (S group, n = 22). Primary end point was postoperative excessive pulmonary flow events (PEPF events) which refers to requirement of cardiopulmonary resuscitation or PDA ligation and/or shunt clipping.
Results: In term of change of preoperative cardiac function by low pulmonary flow strategy, BNP level at operation (213.8 ± 202.8 pg/ml) was significantly reduced compared with that one week before operation (467.5 ± 450.6 pg/ml) (P = 0.005) in S group. Characteristics and perioperative data in detail were shown in table. PEPF events were significantly less frequent in S group. Furthermore, preoperative SpO2, postoperative catecholamine index in S group were significantly lower than that in N group. Preoperative BNP, postoperative maximum lactate level and duration of ICU stay in S group tend to be lower than that in N group.
Conclusions: It is suggested that these strategies contribute to lower postoperative mortality and morbidity of BT shunt for PA or severe PS with PDA-dependent circulation.

Table.

	N group (n = 20)	S group (n = 19)	P value
Body weight (BW) at operation (kg)	3.6 ± 1.5	3.2 ± 0.3	0.18
Age at operation (days)	33.8 ± 11.2	34.5 ± 40.8	0.14
Preoperative SpO_2 (%)	84.8 ± 5.3	80.2 ± 5.8	0.02
Preoperative BNP (pg/ml)	344.1 ± 617.0	213.8 ± 202.8	0.44
Dose of preoperative PD EIII inhibitor at anesthetic induction (μg/Ag/min)	0.02 ± 0.12	0.47 ± 0.13	<0.01
BT shunt graft size/BW (mm/kg)	1.1 ± 0.2	1.1 ± 0.1	0.24
Postoperative excessive pulmonary flow events	6 (30%)	0	<0.01
Hospital death	1 (5.0%)	0	1.0
Postoperative catecholamine index	14.2 ± 7.8	7.8 ± 4.3	<0.01
Postoperative maximum lactate (mmol/L)	6.1 ± 7.6	2.7 ± 1.0	0.06
Duration of intubation time (day)	4.3 ± 14.8	3.1 ± 4.6	0.39
Duration of ICU stay (day)	6.6 ± 5.7	3.9 ± 3.6	0.08

P1423 - NATURAL HISTORY OF PULMONARY TRANSITION IN EXTREMELY PREMATURE INFANTS

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Background/ Hypothesis: Preterm infants are at risk for bronchopulmonary dysplasia (BPD)/death due to pulmonary hypertension (PH). However, normal physiology of pulmonary vascular transition (PVT) after delivery was not studied before. Hence the incidence of PH among extremely preterm infants during the first few days of life is unknown. Our primary objective was to report the temporal profile of PVT in preterm infants. We also hypothesized that infants with delayed PVT will be at higher risk for BPD/death.
Materials & Methods: Prospective cohort study of infants born at <29weeks. Initial echocardiogram was at <48 h of life and repeated every 24-48 h for 14 days. Infants were divided in normal and delayed PVT groups. Resolution of PH at <72 hours of life was considered normal. Delayed PVT was defined as evidence of PH at >72 hours. Recurrent PH after documenting normal pulmonary artery pressure was called early PH. Persistent PH (PPHN) was diagnosed if PH continued till 14 days of life. Incidence of PH was calculated at 0-2, 3-6, 7-10 and 11-14days of life. Multivariable analysis was performed to estimate the risk of BPD/death among infants with delayed PT.
Results: March, 2015-April, 2016, 60 infants were enrolled. The incidence of PH decreased from 71% at 0-2d to 30% at 11-14d of life. Five distinct patterns of PT were recognized i.e. normal PT, normal PT with early PH, delayed PT, delayed PT with early PH and PPHN. Two infants (3%) died before 72 hours. Normal and delayed PT was noted in 26(44%) and 32(53%) infants respectively. Infants with delayed PT were of smaller gestation, lower birth weight and at higher risk for BPD/death (p < 0.005).
Conclusion: Incidence of PH decreases over the 14 days of life. PH can reoccur after normal or delayed PT. Delayed PVT increases the risk for BPD/death in preterm infants.

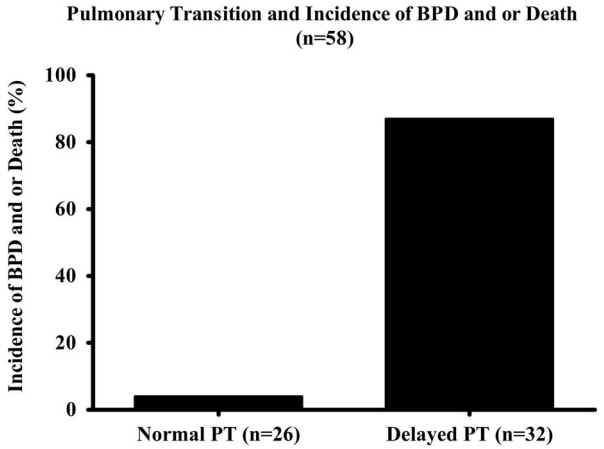

Figure 1.

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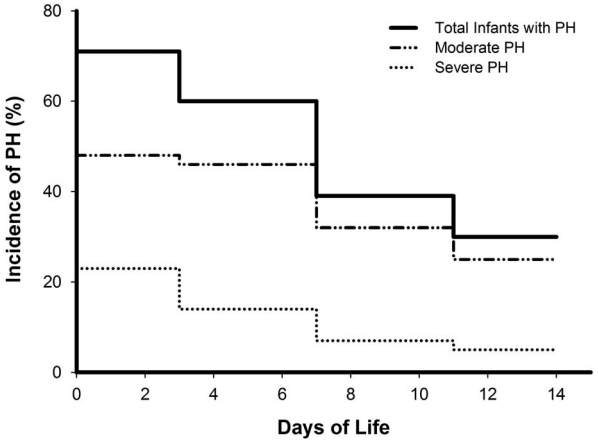

Figure 2.

P1432 - MALNUTRITION IS ASSOCIATED WITH POOR OUTCOMES IN CHILDREN UNDERGOING CONGENITAL HEART SURGERY IN BRAZIL

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Background: Poor nutritional status is associated with adverse outcomes in patients undergoing congenital heart surgery in developed countries. In developing countries, interventions for congenital heart disease are performed late, resulting in a high prevalence of pre-operative malnutrition. Therefore, nutritional status may be an alterable peri-operative factor that may improve surgical outcomes.
Objectives: To assess the nutritional status of children undergoing pediatric congenital heart surgery and its association with clinical outcomes.
Methods: Retrospective review of children undergoing congenital heart surgery at a regional referral center in Brazil. Patients were assessed for nutritional status at admission using World Health Organization weight-for-age standards. The association between nutritional status and clinical outcomes (in-hospital mortality, ICU and hospital length of stay and length of mechanical ventilation) were examined in univariate and multivariate regression models.
Results: Over a two year period, 562 patients underwent corrective or palliative congenital heart surgery. Of these, 217 patients (38.6%) were malnourished. In-hospital mortality was 13.2% (n = 74) with a significantly higher mortality rate among malnourished patients (n = 38, 17.5%) than the non-malnourished patients (n = 36, 10.8%, p = 0.036). Malnourished patients had higher rates of non-cardiac structural anomalies, prematurity, and lower hematocrits. Malnutrition status was also associated with longer post-operative mechanical ventilation times, longer intensive care unit length of stay, and longer post-operative length of stay. In multivariate analyses, malnutrition status was associated with prolonged post-operative mechanical ventilation (additional 36.9 hours, 95% Confidence Interval (CI): 7.1-66.7 hours), increased ICU length of stay (75.7 hours, 95% CI: 28.9-124.1 hours) and increased odds of mortality (Odds ratio: 1.9, 95% CI: 1.06-3.48).
Conclusion: Malnutrition in children with congenital heart disease is associated with poor short-term outcomes after congenital heart surgery. Improvements in nutritional status may improve outcomes.

P1437 - INCOMPLETE AND ATYPICAL KAWASAKI DISEASE IS IT TIME FOR MODIFIED CRITERIA

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Kawasaki disease (KD) lacking full classical criteria is a big diagnostic challenge especially that features are non-specific. We analyzed the data of 4 patients not fulfilling criteria for incomplete KD during same winter season: The first was a 4 months-old boy presenting with generalized mucopapular rash, purulent conjunctivitis, lip fissuring and 1-day of fever. Left coronary system was ectatic initially, progressed to left anterior descending (LAD) aneurysmal dilatation. Symptoms responded to IVIG after an episode of severe anaphylaxis. He continues to have CA aneurysm and is on anticoagulation. Second was a 5 year-old boy manifesting only with 20-days of fever, LAD had fusiform dilatation at its midportion. Fever resolved with IVIG and CA dilatation regressed in 2 months. Third was a 4 year-old boy presenting with 7 days of fever associated with nonpurulant conjunctivitis, follicular tonsillitis, and no other criteria of KD. Both coronary systems were dilated. Responded to IVIG and coronary dilatation regressed within 1 month. Fourth was a 2 year-old boy presenting with 5 days of fever and generalized lymphadenopathy, hepatomegaly, and marked elevation of transaminases. Pericardial effusion was discovered on work-up; on follow-up LAD became ectatic and finger-pealing appeared. Fever responded to IVIG, effusion resolved in 3 days and coronary dilatation regressed in 2 weeks. We report 4 cases diagnosed with KD despite insufficient criteria: typical fever being absent in one, fever was the only manifestation in another, a third with only 2 criteria in addition to fever and a fourth with generalized lymphadenopathy as the predominant feature beside fever. All of which had CA dilatation and responded clinically to IVIG, with the persistence of large coronary aneurysm in one case. We suggest that KD diagnostic criteria be modified to a scoring system including major and minor criteria; incorporating inflammatory markers as a minor criterion.

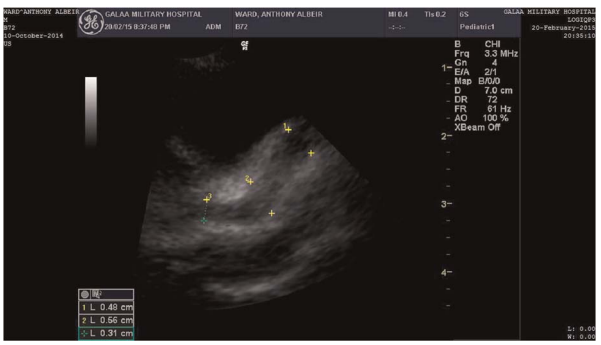

Figure 1.

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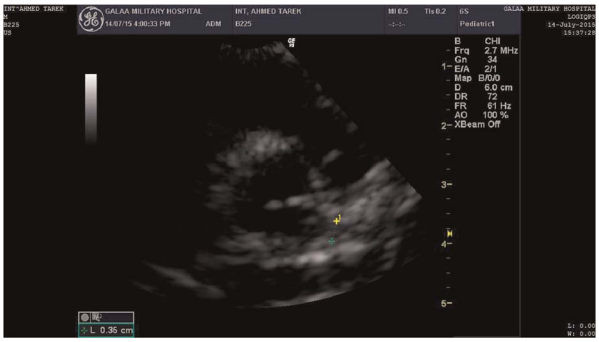

Figure 2.

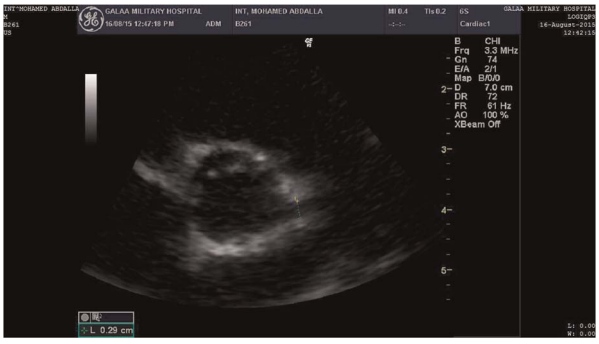

Figure 3.

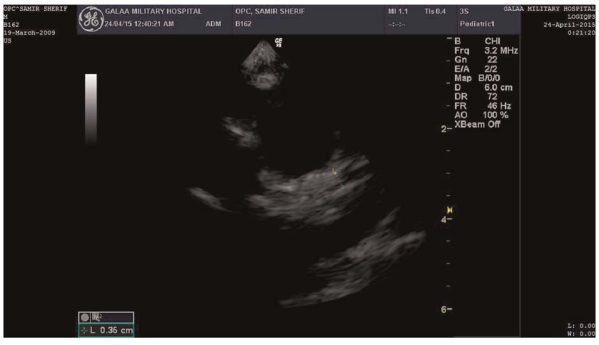

Figure 4.

P1447 - A NOVEL POSTOPERATIVE PAIN CONTROL PROTOCOL TO MANAGE PAIN AND IRRITABILITY IN INFANTS UNDERGOING SUPERIOR CAVOPULMONARY ANASTOMOSIS

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Background: Infants undergoing superior cavopulmonary anastomosis (SCPA) are at risk for immediate post-operative pain. In particular, post-operative headache can be difficult to manage, frequently requiring increasing doses of narcotics to achieve comfort. We describe a novel protocol using a clonidine transdermal patch as an adjunct for pain control after SCPA and its relation to narcotic needs and outcomes.
Methods: Retrospective chart review was performed for all patients who underwent SCPA between 2011 and 2016. Patients were divided into two groups, Group A (no clonidine) and Group B (clonidine). Both groups received acetaminophen and non-steroidal inflammatory drugs every 6 hours as well as narcotics as needed for breakthrough pain. In addition, patients in group B received a 0.05 mg clonidine transdermal patch which was placed immediately after surgery. Timing of extubation, narcotic use within 72 hours of surgery and hospital length of stay were compared between the two groups.
Results: There were a total of 19 cases (Group A n = 9, Group B n = 10). Mean age was 152 days (range 131-1018), mean weight 7.09 kg (range 5.54-11.8). When comparing both groups, patients receiving clonidine (Group B) were extubated earlier (19.9 vs. 59.9 hours, $p < 0.05$). Narcotic administration during the first 72 hours was significantly decreased in patients on the clonidine protocol (3.14 mcg/kg vs. 17.6 mcg/kg of fentanyl, $p = 0.02$) and hospital length of stay was significantly shorter in this group (6.9 vs. 19 days, $p < 0.05$).
Conclusions: Clonidine was a useful adjunct to manage postoperative pain and irritability in this patient population. Narcotic administration was markedly decreased in patients who were on a clonidine patch. Further evaluation of the protocol in a larger patient population is needed. It may prove to be effective in managing post-operative pain and irritability in this patient population.

P1454 - POSTOPERATIVE SECONDARY CHYLOTHORACES ARE ASSOCIATED WITH LYMPHATIC ENDOTHELIAL CELL DEFECTS AND MISEXPRESSION OF BETA ADRENERGIC RECEPTORS

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Background/Hypothesis: Post-operative chylothorax continues to be a significant complication affecting 2-9% pediatric post-cardiac surgery patients. We have shown that lymphatic endothelial cells (LECs) isolated from congenital chylothorax form abnormal lymphatic vessels in a mouse model, overexpress beta-adrenergic receptors (βARs) and respond to propranolol, a βAR inhibitor. As several of the genes necessary for heart development are also required in lymphatic development, we hypothesized that pediatric cardiac patients with postoperative chylous effusion have underlying lymphatic abnormalities predisposing them to the pathology, and that propranolol would improve patient morbidities.
Materials and Methods: Eleven patients with postoperative chylothorax fluid having ≥ 80% lymphocyte count, triglycerides ≥ than serum level, and/or chylomicron positivity were included. LECs isolated from post-operative chylous fluid (n = 8) were subjected to quantitative reverse transcription-polymerase chain reaction to determine expression of endothelial progenitor, LEC and βAR. Human dermal LECs (HdLECs) served as a normal control. Seven patients who failed conservative management consisting of diet modification and diuretics and continued to drain greater than 10cc/kg/day were offered propranolol at 0.5-2.0 mg/kg/day. Fluid output was monitored daily.

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Results: Pleural effusion contained LECs that expressed significantly lower level of LEC genes and high levels of progenitor genes, compared to HdLECs (Table 1). LECs from post-operative chylous effusions significantly overexpressed either β1AR (n = 4/8) or β2AR (n = 4/8). Six of seven patient's fluid volume did not worsen on propranolol, and reduced drainage was observed (Table 2).
Conclusions: Our results suggest that LECs from secondary chylothorax are improperly differentiated and have abnormal beta-adrenergic signaling. Consistent with abnormal beta-adrenergic receptor expression, propranolol treatment correlated with reduced chylous effusions. Thus, propranolol may be efficacious in inhibiting abnormal LEC function and reduce postoperative chylothorax volume, duration, and associated morbidity and mortality.

Table 1. Expression of endothelial progenitor and lymphatic endothelial genes in LECs isolated from lymphatic malformations, postoperative chylothorax and congenital chylothorax relative to HdLECs.

	Endothelial Progenitor Genes				Lymphatic Endothelial Genes			
Anomaly	***Cd34***	***Tie2***	***Vegfr1***	***Vegfr2***	***Prox1***	***Podoplanin***	***Lyve1***	***Ve-cadherin***
Lymphatic Malformation	↑(3/4)	↑(3/4)	↓(5/5)	↓(3/5)	↑(4/5)	↑(5/5)	↓(5/5)	↓(4/5)
Post-op Chylothorax	↑(7/8)	↑(8/8)	n.c.⋆	↑(5/5)	↑(7/8)	↓(8/8)	↓(4/8)	↑(5/5)
Congenital Chylothorax	↑(3/4)	↓(3/4)	↓(4/4)	↓(4/4)	↓(4/4)	↓(5/5)	↓(4/4)	↓(4/4)

⋆n.c. – no change, (n/n) denotes number of population with increased or decreased expression/total populations.

Table 2. Chest-tube drainage in response to propranolol after 12 days.

Patient	Pre-treatment output⋆	Post-treatment output⋆
1	115 mls/d	4 mls/d
2	766 mls/d	82 mls/d
3	172 mls/d	438 mls/d (9 days)⋆⋆
4	678 mls/d	321 mls/d
5	172 mls/d	22 mls/d
6	269 mls/d	164 mls/d
7	645 mls/d	0 mls/d (4 days)⋆⋆⋆

⋆Average chest tube output over 3-day period.
⋆⋆Patient died.
⋆⋆⋆ Drain output was 0 mls on 4th day of treatment.

P1457 - VARIABILITY IN PERIOPERATIVE INOTROPIC USE IN CONGENITAL CARDIAC SURGERY

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Background: Although inotropic support is instrumental in the perioperative care of children undergoing cardiac surgery, there is limited data describing hospital practices and trends.
Objective: We sought to describe perioperative inotrope use, including institutional variability, among US children's hospitals in children undergoing cardiac surgery.
Methods: A multi-institutional, retrospective review of the Pediatric Health Information System (PHIS) identified all hospitalizations involving patients <18y who underwent cardiac surgery and received inotropic support between 2004 and 2015. Inotropic support was defined as preoperative (starting before surgery) or operative (starting on the day of surgery) use of dobutamine, dopamine, epinephrine, or milrinone at the index cardiac surgery. Calcium was not included.
Results: There were a total of 106,921 hospitalizations (median age 0.5y [IQR 0.1-3.2], 55% male, 83% cardiopulmonary bypass [CPB]) across 46 hospitals. Preoperative inotropes were used in 22,301 (21%) hospitalizations, significantly associated with neonatal age (OR 5.6, 95% CI 5.4-5.7), and commonly involved epinephrine (68%) and milrinone (51%). Operative inotropes were utilized in 74,947 (70%) hospitalizations and involved milrinone (68%), epinephrine (66%), dopamine (39%), and dobutamine (24%). Among CPB cases, milrinone was utilized in 74%, followed by epinephrine (69%), dopamine (39%) and dobutamine (25%). Center variability among operative inotropes was wide (milrinone 42-99%, epinephrine 41-100%, dopamine 8-77%, and dobutamine 4-70%). Operative single-drug regimens were used in 25% while 2-drug and 3-drug regimens were used in 40% and 56%, respectively. Common multi-drug regimens were dopamine-epinephrine-milrinone (33%), epinephrine-milrinone (20%) and dobutamine-epinephrine-milrinone (18%). There was significant decline in usage of dobutamine (2%/yr; $r = 0.96$; $p < 0.001$) and dopamine (2%/yr, $r = 0.97$; $p < 0.001$) throughout the study period.
Conclusions: The use of inotropes in children undergoing cardiac surgery widely varies by center. It is unclear if this variation plays any role in differences in outcomes. Further study examining the details of drug administration and association with outcomes may help further improve care.

P1474 - EARLY TREATMENT OF INTRA PERICARDIAL TERATOMA IN A NEWBORN

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Background: Intra-pericardial teratoma (IPT) is a rare congenital condition and well-recognized cause of pericardical effusion (PE), cardiac compression, severe cardio-respiratory distress and hydrops fetalis (HF). Usually, it presents as an intra-thoracic, well-delimited, heterogeneous mass with cystic areas. Most tumors are generally benign, but without an adequate prenatal and postnatal treatment, they can be potentially fatal.
Objective: To presente a case report of successful management of a newborn with an IPT and a large PE diagnosed in the pre-natal period.
Case presentation: A woman was referred at 38 weeks of gestational age (GA) to our service for a large fetal PE and a intrapericardial cystic mass diagnosed at 36 weeks GA by ultrasound (US), with no evidence of HF. Fetal echocardiography showed well-defined intrapericardial tumor, made up by soft tissue mass with cystic components above the root of the great vessels, no structural heart abnormalities and an important PE. There were no other abnormal US findings. An elective cesarean delivery was performed.

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A 3 kg baby girl evolved with important respiratory distress associated with cyanosis soon after birth, requiring tracheal intubation. Postnatal echocardiogram revealed a heterogeneous cystic mass arising from the base of the heart and a massive PE. The child was sent to the operating room for surgical removal of the tumor three hours after birth. Surgical finding showed a mass attached to the lateral wall of the ascending aorta. There was no extension of the tumor into the surrounding structures. The gross specimen was a 5 × 5 cm mass and histopathological exam confirmed the diagnosis. The postoperative course was uneventful and she was discharged from the hospital one week later.
Conclusion: Early surgery may be a successful option for newborn who present with important pericardial effusion and respiratory distress, with an excellent result.

P1492 - EARLY OUTCOME AFTER REPAIR OF TETRALOGY OF FALLOT IN ADULTHOOD IN THE CURRENT ERA

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Background: Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease. Primary surgical repair is usually performed in early childhood with excellent results. However, in developing countries we still encounter adults with uncorrected TOF or whom only had a palliative procedure in early childhood. Repair of TOF in adulthood has been associated with prolonged intensive care stay related to myocardial dysfunction,arrhythmia and end-organ injury.
Methods and Patients: Retrospective study of all patients >16 years who underwent primary repair of TOF in our Centre since 2013. There were 29 patients (18 males) including 4 with a palliative shunt. Median age was 21 years (16 to 41 years) and percutaneous oxygen saturation 80% (45 to 98%) with saturation <75% in 13 patients. Median haematocrit value was 56% (33 to 73%). Surgical technique involved transatrial/transpulmonary repair with patch augmentation of right ventricular (RV) infundibulum in cases where the pulmonary valve remained functional (16 patients) or placement of a valved RV to pulmonary artery conduit if the valve was inadequate (13 patients). Milrinone and Norepinephrine were routinely used for post-operative inotropic support.
Results: There were no operative deaths. twenty seven patients were extubated within 36 hours, 2 required re-ventilation due to lung reperfusion injury and a further 2 were ventilated for 5 days due mediastinal bleeding and aspiration, respectively. Four Patients developed pleural effusion necessitating drainage, and 2 suffered a neurological complication (stroke and seizures) with full recovery. Four patients had 2nd degree heart block with no need for pacemaker, and none suffered junctional ectopic tachycardia. No patient required renal dialysis. Median hospital stay was 8 days (5 to 61 days).
Conclusion: In this contemporary series, repair of TOF in adulthood was associated with excellent early post-operative survival and acceptable morbidity.

P1498 - PERIPHERALLY INSERTED CENTRAL VENOUS CATHETERS IN CHILDREN WITH HEART DISEASE FIRST EXPERIENCE

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Background: In recent years the use of peripherally inserted central venous catheter (PICC) is significantly increasing and this increase has led to the expansion of indications such as pediatric intensive care. Children with congenital heart disease are a specific patient population in which the venous system thrombosis and its sequelae significantly contribute to morbidity and mortality. Prevalence and severity of thrombosis is associated with unique factors such as developmental and acquired bleeding disorders, complex perioperative care requiring central venous access and catheterization procedures.
Material and Methods: The aim of this study was to evaluate the initial experiences with the PICC at CICU in Pediatric Cardiac Centre in Bratislava from December 2015 to November 2016. Data collection was performed prospectively in patients who have undergone the PICC placement procedure.
Results: During the review period 42 lines were inserted in 34 patients. The median age at the time of insertion was 6.3 months (1 day - 21 years), median weight was 5 kg (2.4-90 kg). The PICC insertion was ultrasound guided through the basilic, cephalic, axillary, and femoral vein in 13 (31%), 4 (9.5%), 6(14.3%), 29 (45.2%) cases respectively. The PICC lines used were 1.9 Fr single lumen, 2.6 Fr double lumen and 4Fr single and double lumen used in 2, 35, and 3 patients respectively. The main indications were intensive complex therapy, parenteral nutrition and antibiotics therapy in 31 (73.8%), 4 (9.5%), and 7 (16.7%) patients respectively. The median duration of PICC was 22 days (14-98days). The complication rate was 9.5% including 3 line fractures, 1 infection at the insertion site and 1 venous thrombosis.
Conclusion: We believe based on our initial experience that the use of PICC in pediatric patients with heart diseases in the intensive care settings is a possible solution for central venous access.

P1503 - DECREASED OPIATE USAGE BY INCORPORATING OFIRIMEV IN POST OPERATIVE PAIN MANAGEMENT IN CONGENITAL HEART SURGERY

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Background: Opioids and benzodiazepines are frequently used to manage post-operative pain and sedation in patients undergoing congenital heart surgery. In recent years, Ofirmev has been incorporated into the post-operative pain management plan.
Methods: We reviewed the use of opioids and benzodiazepines in two cohorts from our congenital cardiac surgery program. Group 1 consisted of congenital heart cases performed between 2012-2014. Group 2 were cases performed between 2014-2016. Data was extracted from Vizient, which comes from UHC institutions, and specifically looked at the type of medication used, percent of cases requiring the use of opiates, benzodiazepines and Ofirimev and number of treatment days each case required. A comparison of overall opiate, benzodiazepine and Ofirimev utilization between the two cohorts was completed.
Results: 37 congenital heart surgeries occurred between 2012-2014. Between 2014-2016, 173 heart surgeries were performed. 92% of cases from both cohorts received opiates post-operatively while 68% of Group 1 received benzodiazepines, compared to 69% of Group 2. Ofirimev was not used in post-operative management for cases performed between 2012-2014, whereas, 51% of cases from 2014-2016 received Ofirimev. With the addition of

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Ofirimev in post-operative pain management, there was an overall decrease in length of opiate treatment with the current cohort receiving 3.8 days of opiates compared to the historical group at 7.7 days. Benzodiazepine use was 3.2 days in the recent cohort compared to 3.8 days in the previous cohort.
Conclusions: When compared to the historical cohort, patients undergoing heart surgery between 2014-2016 received fewer days of opiates and more Ofirimev. Overall use of benzodiazepines was similar for each group. Although early results suggest that Ofirimev decreases opiate and benzodiazepine usage in post-operative pain management, further studies are needed.

P1506 - SIGNIFICANTLY REDUCING BLOOD DRAWS IN THE CONGENITAL CARDIAC INTENSIVE CARE UNIT BY MULTI DISCIPLINARY TEAM APPROACH AND GOAL ALIGNMENT

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Background: Communication is critical and is important the entire team understands the goals for the patient. Blood draws are a leading cause of unnecessary harm to the patient from (1) requirement for being stuck for the blood draw, (2) cost, and (3) iatrogenic blood loss. Volume of blood per test is >5 ml, traditional, >2 ml, micro-tubes, and 100 microliters, point of care testing (POC).
Methods: We looked at all procedures that required cardiopulmonary bypass from November 2013 to November 2016 program, after establishing a new congenital heart program, with the crucial goal of decreasing the overall impact to our patients. We developed a multi-disciplinary team to (1) reduce blood draws, (2) use POC as the primary lab, (3) reduce waste of blood, and (4) develop standard protocols for blood draws.
Results: We performed eighty surgeries. In the traditional ICU there was a median POC of 15 tests, median CBCs of 7 tests and median chemistries of 9 tests, and for the new congenital cardiac ICU there was a median POC tests of 7, median CBCs of 1 test, and median chemistries of 1 test. In attributed blood loss for tests in the traditional ICU on average was 267 ml in FY14, 124 ml in FY15 and 92 ml in FY 16, for the congenital cardiac ICU was 7.3 ml in FY 14, 3.6 ml in FY 15, and 2.4 ml in FY 16, all significantly less, $p < 0.05$.
Conclusions: We developed a multi-disciplinary team to reduce the blood draws, and developed a method to reduce blood waste during the blood tests. Using a 2-stopcock technique, with the waste blood remaining inline and be given back immediately without disconnecting. We could move to a POC primary blood draw system that significantly reduced blood draws and blood loss.

P1507 - SIMULATION TRAINING ENGENDERS HIGHER COMFORT LEVELS AMONG CAREGIVERS AND LOWERS TIME TO RESPONSE IN LOW RESOURCE INTENSIVE CARE UNIT

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Background/Hypothesis: Although simulation-based Crisis Resource Management (CRM) training has demonstrated improvement in team performance in many high-resource cardiac intensive care (CICU) environments, its impact in low resource CICUs has not been studied. Endemic to such environments are multifactorial barriers that inhibit effective nurse-physician communication in crisis situations. We hypothesize that CRM training leads to improvements in team dynamics and time to appropriate medical response in such settings.
Materials and Methods: In this prospective observational study, the effects of CRM training on team dynamics and performance were assessed in 23 healthcare providers at a single CICU in South-East Asia. A 5-day training program was utilized consisting of didactic sessions and CRM training exercises. Improvements in team dynamic (effective and closed-loop communication, role delegation, idea acceptance) were assessed using participant questionnaires, expert evaluations, and video analysis. The time to appropriate medical intervention following a simulated perturbation in vital signs (fluid bolus, inotropic support, bag-mask ventilation) was measured over the training period.
Results: Through questionnaires, participants noted significant ($P < 0.05$) improvement in team dynamics and performance over the training period (Table 1). Independent external observers also reported improvements in team performance. Blinded video analysis of training exercises revealed significant increase in frequency of closed-loop communication between caregivers (2.2/min ± 0.02 vs. 4.4/min ± 0.6, $P < 0.05$), and a trend towards decrease in times to appropriate clinical intervention (Table 1). Through questionnaires sent 30 days following CRM training, caregivers expressed improved communication within their native clinical environment as a result of CRM training.
Conclusions: The study demonstrates the feasibility and effectiveness of CRM training at improving team dynamics, particularly between nurses and physicians, in global health environments, indicating its potential role as a training tool in such settings.

Table 1.

Component Assessed	Day #1 Median Score (IQR) N = 15	Day #2 Median Score (IQR) N = 7	Day #3 Median Score (IQR) N = 7	P-Value⋆
You understood your role#	1 (1-2)	1 (1-1)	1 (1-1)	0.043
Others understood roles#	2 (2-3)	1.5 (1-2)	1 (1-2)	0.026
Problem presentation#	2 (1-2)	1 (1-1)	1 (1-1)	0.004
Communication Thresholds#	1 (1-2)	1 (1-1)	1 (1-1)	0.020
Overall communication#	2 (1-3)	1 (1-2)	1 (1-1)	0.018
Ideas valued#	2 (1-2)	2 (1-2)	1 (1-1)	0.033
Mean Time to Intervention for Hypotension	69.0	44.5	26.5	—
Mean Time to Intervention – Bag and Mask Ventilation	90.0	71.0	27.0	—

#Responses to participant questionnaires.
⋆Kruskal Wallis statistical analysis; IQR – interquartile range; 1-strongly agree/excellent, 2-agree/good, 3-neutral/fair, 4-disagree/poor, 5-strongly disagree/very poor.

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P1510 - ACUTE KIDNEY INJURY SCORING SYSTEM IS A BETTER PREDICTOR OF INCREASED LENGTH COMPARED TO INOTROPE SCORE

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Background: Acute Kidney Injury (AKI) and increased inotrope score have been shown to be associated with increased morbidity and mortality in pediatric congenital cardiac surgery. AKI has been shown to occur in adult and pediatric post-operative cardiac surgical patients between 18 and 60%. The purpose of this study was to evaluate the incidence of AKI comparing the pediatric risk, injury, failure, loss, end (pRIFLE) and the acute kidney injury network (AKIN) scoring systems to inotrope score and post-operative lactate.
Methods: Retrospective analysis of all congenital heart surgeries requiring cardiopulmonary bypass between 2014 and 2016. All patients received aggressive dilutional ultrafiltration on cardiopulmonary bypass. All neonates received a peritoneal catheter for fluid removal only, no dialysis was used. Values for serum creatinine measured pre-operative (baseline), first on arrival to the intensive care unit (ICU), twenty-four, and forty-eight hours, urine output for the first 24 hours, lactate, bypass time, ventilation time, inotrope scores, and length of stay (LOS) were pulled from the quality database.
Results: Eighty cardiopulmonary bypass cases were performed, but 4 cases were excluded for pre-operative renal failure leaving seventy-six bypass cases. AKI was associated with significantly increased CPB time (No AKI at 134 min AKIN and 130 min pRIFLE vs any AKI 191 min AKIN and 198 min pRIFLE), increased creatinine (AKIN 270%, pRIFLE 235% over baseline), greater reintubation rate(2-5% versus 25-36%), and LOS (No AKI 10 days versus AKI 36 days AKIN and 25 days pRIFLE). There was not a significant difference in peak inotrope score over 48 hours post-op or peak lactate post-op.
Conclusion: The incidence of AKI significantly increases post-operative length of stay. Inotrope score, while important, does not necessarily lead to improvement in length of stay. Focus on fluid management peri-operatively, and specifically fluid overload, may lead to decreased length of stay.

P1514 - FONTAN COMPLETION DURING WINTER SEASON IS NOT ASSOCIATED WITH SIGNIFICANTLY HIGHER MORTALITY OR SEVERE MORBIDITY IN THE EARLY POSTOPERATIVE PERIOD

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Objectives: The aim of our study was to compare postoperative outcomes after total cavopulmonary connection (TCPC) between patients who were operated during winter and summer season.
Methods: We retrospectively studied 193 patients who underwent extracardiac TCPC completion at our institution between 1995 and 2015. 63 (33%) patients were operated during winter (november-to-march) and 130 (67%) patients during summer season (april-to-october). We compared the following post-operative outcomes: early mortality (i.e. within 30 days postoperatively), need for Fontan takedown, intubation time (hours), incidence of fast track extubation (extubation within 6 hours postoperatively), incidence of pleural effusions longer than 10 days, incidence of low cardiac output (i.e. need for intravenous catecholamine therapy longer than 72 hours), need for dialysis, intensive care unit (ICU) and hospital stay (days). Data are presented as percentage or median values, respectively.
Results: Intubation time (13 vs. 11.5 hours, $p = 0.047$) was slightly longer in patients operated during winter season, however, early mortality (8% vs. 5%, $p = 0.53$), need for Fontan takedown (2% vs. 1%, $p = 0.55$), low cardiac output (19% vs. 9%, $p = 0.09$), dialysis (13% vs. 8%, $p = 0.4$), fast track extubation (13% vs 18%, $p = 0.5$), prolonged pleural effusions (36% vs. 29%, $p = 0.4$), ICU (4 vs. 3 days, $p = 0.2$) and hospital stay (15 vs. 14, $p = 0.05$) were not significantly different between both groups.
Conclusion: TCPC completion during winter season was not associated with higher mortality or severe morbidity in the early postoperative period. These results suggest that TCPC completion during winter season might be performed at no significant additional risk for patients.

P1529 - ASSESSMENT OF SERUM VITAMIN A LEVELS IN INFANTS WITH CONGENITAL HEART DISEASE

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Background: Prevention of congenital heart disease (CHD) has been hampered by a lack of information about modifiable risk factors for abnormalities in cardiac development. Vitamin A plays an important role in the periods of rapid cellular growth and differentiation, especially during pregnancy. The aim of this study is evaluation of vitamin A serum levels in children with CHD and compare to children without CHD.
Materials and Methods: In a case-control study that was conducted in 2015 in Mashhad /Iran, serum levels of vitamin A in 30 children with CHD were compared to 30 controls. Cases of congenital heart disease diagnosed by echocardiography and were recruited by convenience sampling. The control group was selected from infants who referred to health centers for routine care. Exclusion criteria for both groups were concurrent systemic diseases and malnutrition. Data analysis was done in SPSS V 20 software and descriptive statistics, t-test was used.
Results: The mean age in cases was 10.25 ± 6.5 days and in controls was 18 ± 6 days. In both groups, 18 patients (60%) were male and 12 patients (40%) were female. In CHD patients, 10 cases (33.3%) had cyanotic heart disease and 20 cases (66.7%) had acyanotic heart disease. The mean serum vitamin A values in subjects 11.66 ± 13.58 ng/dl and controls 22.54 ± 16.3 ng/dl was significantly different, ($P < 0.05$).
Conclusions: There was a significant difference in serum vitamin A values in subjects and controls. Therefore, we suggest that the serum levels of vitamin A in children with CHD should be checked routinely.

P1544 - THE CURRENT STATE OF SURGICAL INTERVENTION FOR CONGENITAL HEART DISEASE IN TRISOMY 13 AND TRISOMY 18

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Introduction: Trisomy 13 (T13) and Trisomy 18 (T18) are relatively common chromosomal disorders often observed in pediatrics practice. Both T13 and T18 are highly associated with congenital heart disease (CHD). For a long-time, aggressive surgical intervention to CHD accompanied with 13 T and 18 T were refrained due to its extremely poor track records, however, in some advanced hospitals, owing to recent advancements, surgical intervention is being considered and has resulted in stabilizing general conditions. Having said, it is yet uncertain whether aggressive surgical interventions have brought about a truly positive impact. Under such circumstances, we have reviewed our experience of surgical interventions for CHD in T13, T18 retrospectively to clarify its effectiveness.
Subjects and Results: Thirty consecutive patients of T13 and T18 were enrolled. Twenty-eight of the patients had CHD, and 7 of those underwent surgical intervention. Performed procedures were palliative operation for all cases. During the study period, only 2 patients survived; of these, 1 underwent surgery and the other did not. Of those 28 patients, median survival days of patients who underwent surgery was 443 days while median survival days of those who did not had surgical intervention was 94 days. However, only 2 of 7 patients (28.5%) who underwent surgery were able to discharge from the Neonatal Intensive Care Unit (NICU) while 9 of the 23 patients (39.0%) who did not undergo surgery were able to discharge from the NICU.
Discussion: Judging from our experiences, it seems that surgical intervention to CHD either T13 or T18 prolongs lifespan. However, even if we did surgical interventions, few of those patients were discharged from NICU and most of those forced to continue intensive care. Considering our results, even if we could prolong lifespan by surgical intervention, we are not sure if it is truly beneficial.

P1546 - EFFECTIVENESS OF INFLIXIMAB IN REFRACTORY KAWASAKI DISEASE

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Background: Infliximab (IFX) is recognized as an additional therapy for Kawasaki disease (KD). We examined the effectiveness of IFX therapy in refractory KD.
Materials and Methods: Between 2013 and 2016, 11 patients with refractory KD including 1 recurrent case were enrolled. All patients were primary treated with intravenous immunoglobulin (IVIG) at single dose of 2 g/kg. Refractory to IVIG was defined as persistent fever over 37.5 degrees Celsius 48 hours after IVIG. IFX was administered as a second or third line treatment at 5 mg/kg intravenous infusion. IFX was considered to be effective when body temperature remained under 37.5 degrees Celsius 48 hours after administration.
Results: IFX was administered as second line treatment in 4 patients, and third in 7 patients. Median age was 33 months (range: 9-85 months), median interval from the onset to initial IVIG was 4 days (range: 3-5 days), median interval from the onset to IFX administration was 9 days (range: 6-20 days), median interval from the onset to removal of fever was 12 days (range: 8-21 days), and median hospitalization was 20 days (range: 10-59 days). Initial laboratory examinations showed elevated C-reactive protein (median 9.3 mg/dL: range 4.25-20.8 mg/dL) and neutrophil counts (median 11.0 ×109/L: range 5.5-20.5 ×109/L). At 48 hours after IFX infusion, C-reactive protein decreased in 9 patients, whereas neutrophil counts decreased in 6 patients. IFX was effective in 8 patients, 6 of them brought a high temperature down within 24 hours after IFX infusion. Three non-responders were treated with plasma exchange. Coronary aneurysms at discharge from the hospital were detected in 3 patients. No severe adverse effect was attributable to IFX.
Conclusions: IFX would be provided safely in pediatric patients with KD. Our results suggest that the further evaluation and accumulation of evidence in refractory KD regarding IFX treatment is necessary.

P1551 - COMPLICATIONS OF TAKEUCHI'S SURGERY ANEURISM OF PULMONARY TRUNK CASE REPORT

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Introduction: The anomalous origin of the left coronary artery of the pulmonary trunk (ALCAPA) is the most frequent congenital coronary anomaly and represents one of the most common causes of ischemia and myocardial infarction in children Echocardiography is the most commonly used method for the diagnosis of ALCAPA, Cardiac catheterization with coronary angiography is the exam of choice to confirm the diagnosis and should be used whenever echocardiography leaves doubts. The treatment of ALCAPA is always surgical. One of the most commonly used techniques is the Takeuchi procedure. The complications most associated with this technique are pulmonary supravalvular stenosis, coronary-pulmonary fistula and aortic valve insufficiency.
Case Description: ESS, 5 years and 6 months, male, diagnosed at 3 months of age, of anomalous left pulmonary trunk coronary artery (ALCAPA), complicated with significant mitral regurgitation (IM) and significant left ventricular systolic dysfunction, submitted to Takeuchi's procedure. After surgery, he had a progressive improvement in the left ventricular ejection fraction (LVEF). It evolved with turbulent flow in the pulmonary artery and progressive increase of the supravalvular pulmonary gradient (Gd) on subsequent echocardiograms and decrease of LVEF as times go on. He underwent thoracic angiotomography 1 year and 11 months after surgery, showing pulmonary trunk stenosis and left coronary redirect tunnel obstruction, as well as an image suggestive of myocardial infarction in the anterior and mid-apical segments of the LV.He performed a new cardiac catheterization 5 years after the surgical procedure with a description of normal LVEF, important pulmonary supravalvular stenosis near the

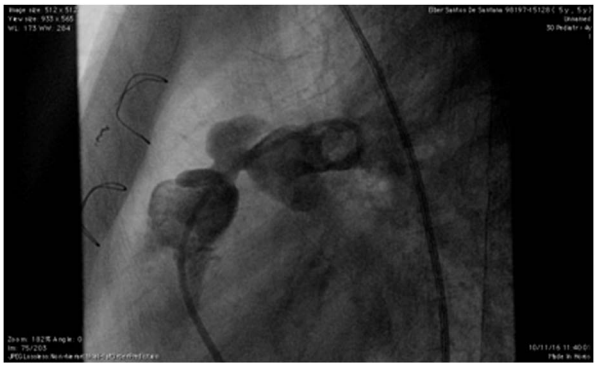

Figure 1.

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pulmonary artery bifurcation with Gd 80 mmHg to the method (Figure 1). Additionally, visualized in profile image suggestive of aneurysm of the pulmonary trunk at the stenosis level (Figure 2).
Discussion: ALCAPA is a pathology with high mortality and morbidity,The case in question brings pulmonary trunk aneurysm as a possible complication associated with the technique.

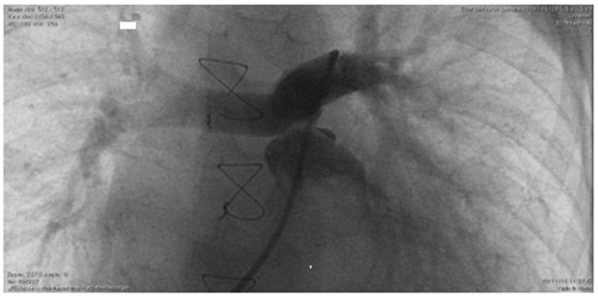

Figure 2.

P1554 - THE MULTIDISIPLINARY EXPERIENCE OF MOVING FROM A TRAINEE NURSE PRACTITIONER ROLE TO ADVANCED NURSE PRACTITIONER ROLE ON PAEDIATRIC CARDIAC INTENSIVE CARE

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Background: Advanced nurse practitioner roles have developed from a perceived need within the NHS and worldwide. The success of Advanced nurse practitioners is widely documented in primary care and adult services while there is only a small amount of literature within paediatrics and less again within specialty services, such as paediatric intensive care. The study aim was to improve the trainee role as it moves into an advanced nurse practitioner role through evaluation of the current trainee nurse practitioners on paediatric cardiac intensive care and a comparison to the established advanced nurse practitioners within children's acute transport service.
Methods: The research was a pilot study which utilized qualitative questionnaire. Questionnaires were distributed within the multidisciplinary teams on cardiac intensive care and the children's acute transport service using an online application. Sampling was purposeful to include a representative sample of the multidisciplinary teams. Findings: Content analysis was used to identify themes from the research. Six themes emerged; positive impact on patient and staff experience, provision of expert care, recognition of blurring of boundaries and unclear role definition, difference in working across the two studied specialties, development suggestions for a more effective advanced nurse practitioner role on cardiac intensive care and perceived impact of an advanced nurse practitioner program on nursing.
Conclusions: Having a trainee nurse practitioner role on cardiac intensive care has been identified by the multidisciplinary team as a positive experience for staff, patient and families. There is an opportunity to develop and grow the advanced nurse practitioner program on cardiac intensive care. It is important further research is undertaken into the scope and practice of advanced nurse practitioners to allow greater understanding of their impact in paediatric intensive care.

P1576 - TRANSCIENT ISOLATED SEPTAL AND RIGHT VENTRICULAR HYPERTROPHIC CARDIOMYOPATHY IN A NEONATE

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Hypertrophic cardiomyopathy usually involves left ventricle. Isolated involvement of the septum and right ventricle without involvement of left ventricle in the neonatal period is a very rare condition. Clinical manifestations can be serious and 25% can develop signs of heart failure usually secondary to obstruction of the right outflow tract. Early diagnosis is vital since this condition may be reversible with appropriate treatment. We report the case of a male newborn who develop cyanosis in his first hour of life. He was born via cesarean section due to perinatal stress. There was no history of maternal or fetal metabolic disease, no gestational diabetes mellitus. Pregnancy was eventful due to a maternal left mastoiditis which required mastoidectomy and antibiotic and analgesic therapy (NSAID) at 34 gestational week. Normal birth weight (3.3 kilogram), APGAR score at 1 and 5 minute was 7 and 9 respectively. Oxygen saturation on room air was 65%, and was rised to 85% with oxygen supplement. A grade II systolic murmur was present in the tricuspid focus. ANP values were high. Echocardiogram revealed massive hypertrophy in the right ventricle with septal and right ventricular hypertrophic cardiomyopathy, with a very restricted diastolic cavity and a right to left shunt across de patent foramen ovale, grade II tricuspid regurgitation which estimated a right ventricular pressure of 50 mmHg. Patient was admitted to neonatal intensive care unit and put on phosphodiesterase 3 inhibitor and sildenafil to decrease pulmonary vascular resistance. An oxygen saturation of 90% was achieved with the supplementation of 40% oxygen. Saturation values rose up to 95% as of day three and oxygen supplement weaned. The patient was discharged oxygen-free and breastfed on the 10th day. With clinical and echocardiographic follow up patient continuos with normal right ventricular and saturation values.

Table.

	03.09.16 0 day	03.11.16 2 days	03.15.16 6 days	05.15.16 1m6d	09.02.16 5m24d
LVEDD (MM)	14.6 (zs -2.2)	14.7 (zs -2.2)	16 (zs -1.19)	20 (zs +0.13)	25.8 (zs -0.09)
LVESD (MM)	8 (zs -2.25)	8.5 (zs -2.2)	10 (zs -0.86)	12.8 (zs +0.37)	17.1 (zs +0.49)
IVSD (MM)	8 (zs +3.8)	7.7 (zs +3.5)	5.3 (zs +1.8)	5 (zs +1.32)	5.2 (zs +0.85)
LVPWD (MM)	10.4 (zs +4.2)	9.1 (zs +3.48)	6.7 (zs +1.88)	7 (zs +1.8)	5 (zs +1.64)
TAPSE (MM)	7 (zs -1.7)	7.1 (zs -1.71)	7 (zs +1.27)	15 (zs +1.27)	15 (zs +1.27)
EF (%)	66	75	69	71	74
SF (%)	33	40	35	36	41

P1594 - URINARY SYNDECAN 1 IMPROVES SEVERE ACUTE KIDNEY INJURY PREDICTION AFTER PEDIATRIC CARDIAC SURGERY

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Introduction: Acute kidney injury (AKI) is a common occurrence after pediatric cardiac surgery and is associated with adverse patient outcomes. Previous we have demonstrated plasma syndecan-1 (a biomarker of endothelial glycocalyx damage) is associated with cardiac surgery-associated AKI. Here, we evaluated the value of urinary syndecan-1 in predicting AKI in this population.
Methods: Prospective cohort study with 83 patients under 18 years old submitted to cardiac surgery at one reference institution. Postoperative urinary syndecan-1 within the first 2 hours after cardiac surgery was collected and normalized to urinary creatinine. Severe AKI - defined according KDIGO stage 2 or 3 was the main outcome. We evaluated if urinary syndecan-1 was independently associated with severe AKI and if it adds prognostic value when added to a clinical model (including age, sex, CPB use, CPB time more than 120 minutes, RACHS-1 3 or more, pre- operative eGFR percentile, percentile of systolic blood pressure at ICU admission, maximum vasoactive inotropic score in the first 48 hours and lactate at ICU admission) performance to predict severe AKI.
Results: Urinary syndecan-1 levels in the postoperative period was higher in patients developing severe AKI (1,062 vs 272 ng/mg-Cr, $p < 0.001$). After adjusting for several confounders, urinary syndecan-1 remained independently associated with severe AKI (OR 1.02 95% CI 1.01-1.04 for each 10ng/mg-Cr increment). The accuracy of postoperative syndecan-1 for diagnosis of severe AKI was moderate (AUC-ROC of 0.78 95% CI 0.58-0.98). The addition of urinary syndecan-1 improved the discrimination capacity of a clinical model (from 0.81 to 0.89, $p = 0.008$).
Conclusions: Postoperative urinary syndecan-1 is associated with subsequent severe AKI and adds predictor value to a clinical model in predicting severe AKI after pediatric cardiac surgery. It may be useful for risk stratifying patients undergoing cardiac surgery.

P1597 - CLINICAL PROFILE AND TREATMENT OUTCOME OF BLOOD CULTURE NEGATIVE (BCN IE) INFECTIVE ENDOCARDITIS IN CHILDREN A TEN YEAR RETROSPECTIVE STUDY

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Background: Infective endocarditis (IE) remains to be a dreaded complication of congenital and rheumatic heart disease in a third world country where corrective surgical intervention is not readily accessible and affordable. The higher incidence of BCN-IE, whether due to fastidious organisms or technical inadequacies in a government tertiary center, makes it more challenging to manage these cases. Evaluation of clinical practice and outcomes amidst international guidelines on BCN-IE is necessary We conducted a ten-year retrospective study of BCN-IE in children admitted at the University of the Philippines-Philippine General Hospital (UP-PGH) to describe their clinical profile, echocardiographic findings, and antibiotic treatment; and correlate with clinical outcome.
Materials and Methods: Demographic data, clinical presentation, previous antibiotic use, echocardiographic findings, and antibiotic treatment of children with BCN-IE admitted at UP-PGH from 2004-2013 were recorded and analyzed. The clinical outcome of patients, whether favorable (discharged) or unfavorable (died, went home against medical advice, re-admitted one month after discharge) were likewise noted. The Fischer's exact test was used to evaluate association of outcome and clinical features, echocardiographic findings and antibiotic regimen.
Results: Among 91 patients diagnosed with infective endocarditis, 61.54% had BCN-IE. Age ranged from 2 weeks to 18 years old (mean = 10.15 years). Thirty four (52.86%) patients had congenital heart disease in which VSD (25%) was the most common. Nineteen (33.93%) had Rheumatic Heart Disease. Clinical presentation and echocardiographic findings were not associated with outcome. The use of penicillin G and amikacin is associated with unfavorable outcome (11 out of 38, with 5 deaths) ($p = 0.007$, $\alpha = 0.05$).
Conclusion: The trend to unfavorable outcome with use of penicillin suggests adhering to international guidelines for antibiotics of choice for BCN-IE with need for re-evaluation.

P1603 - FUTILITY OF ROUTINE METHEMOGLOBIN LABORATORY ASSAYS IN CRITICALLY ILL PEDIATRIC PATIENTS RECEIVING INHALED NITRIC OXIDE

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Background: Inhaled nitric oxide (iNO) is metabolized to methemoglobin and has historically resulted in methemoglobin levels of up to 10.4% in neonates. Methemoglobinemia above 3% related to iNO administration has been linked to increased mortality in some patient populations. It is a standard of care at our institution to measure methemoglobin concentrations every 24 hours while receiving iNO therapy. We aimed to determine the incidence of methemoglobinemia in pediatric patients receiving inhaled nitric oxide.
Methods: A retrospective chart review was performed from 10/18/2014 to 11/18/2016 at our institution for all pediatric patients less than 18 years of age receiving iNO therapy. Patients receiving iNO with at least one methemoglobin concentration were included in the analysis. Patients with methemoglobinemia on admission as well as those with an underlying risk factor for methemoglobinemia were excluded from the analysis. Data are represented as mean (standard deviation), median (interquartile range), or absolute counts (%) as appropriate.
Results: During the study period 1702 methemoglobin concentrations were collected in 248 patients. Median age and weight were 0.33 years (0.04–0.83) and 4.6 kg (3.38–8.00), respectively. Fifty percent of patients were admitted to the cardiac intensive care unit followed by 25% admitted to both the pediatric and neonatal intensive care units. The mean methemoglobin concentration was 1.33% (+/-0.42) while receiving a mean iNO dose of 11.71 ppm (+/-7.97). Twenty-nine patients had a total of 131 methemoglobin concentrations analyzed while receiving iNO doses greater than 20 ppm. The mean methemoglobin concentration in these patients was the same as the entire cohort at 1.33% (+/-0.42).
Conclusions: Pediatric patients receiving inhaled nitric oxide at doses less than 40 ppm have a low risk of developing methemoglobinemia. Methemoglobin monitoring in pediatric patients receiving iNO therapy should be based upon clinical evidence. Routine monitoring is unnecessary and should be avoided.

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P1632 - EARLY LONGITUDINAL NEURODEVELOPMENTAL QUALITY OF LIFE AND FAMILY IMPACT OUTCOMES FOR INFANT SURVIVORS OF EXTRACORPOREAL LIFE SUPPORT

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Background: Extracorporeal Life Support (ECLS) treatment is well-established in neonatal and paediatric intensive care. International data pooling and analysis has significantly contributed to improved outcomes and understanding of co-morbidities, including brain injury, following ECLS. However, there is little longitudinal data describing functional health and family impact of infant survivors.
Methods: Infants receiving ECLS from 2013 to 2015 underwent sequential assessments. Demographic, clinical data, Bayley Scales of Infant Development, Health-Related Quality of Life (HRQoL) scores, and Parenting Stress (PS) indices were collected longitudinally. Enrolment and assessment of infants is ongoing.
Results: Infants were assessed at 4-6 (N = 17), 12 (N = 13), and 24 months (N = 3) of age. Median age at cannulation was 17 days (IQR = 49), weight 3.9 kg (IQR = 0.86), and duration of ECLS 51 hours (IQR = 136). Reasons for cannulation were cardiac failure (N = 7), respiratory failure (N = 6) and Extra-Corporeal Cardiopulmonary Resuscitation (ECPR) (N = 4). Veno-arterial cannulation occurred centrally (N = 8) and peripherally (N = 9). Average range (±1 SD) for language and cognitive domains were observed at 6 (92.7 ± 12.0; 90.3 ± 9.9) and 12 (92.9 ± 12.9; 93.8 ± 15.0), but low average range (1-2 SD) at 24 months (84.3 ± 22.7; 85.0 ± 13.2), as were motor scores at all age points (82.5 ± 18.7; 83.6 ± 13.4; 88.3 ± 28.9). Average range for social-emotional and adaptive behaviour domains were recorded at 6 (94.0 ± 28.8; 79.3 ± 33.9) and 12 (101.1 ± 16.2; 86.8 ± 13.1), but well below normal (<2 SD) at 24 months (61.7 ± 54.0; 47.0 ± 42.0). Scores in ECPR were generally higher compared to non-ECPR, but in average range across all domains at 6 and 12 months (see Table). Parent-reported HRQoL was significantly impaired (<2 SD) at 6 and 12 months, though normal at 2 years. PS was normal at all time points.
Conclusions: Early results and small numbers indicate that infant survivors bear functional health and family impact deficits over the 2 years following ECLS, particularly in motor, social-emotional, behaviour and HRQoL domains, and that ECPR may not make them worse.

Table.

	ECPR		Non-ECPR	
Bayley Scales of Infant Development 3rd Edition	**6 Months N = 4**	**12 Months N = 2**	**6 Months N = 13**	**12 Months N = 11**
Cognitive Skills (mean ± SD)	90 ± 7.1	103 ± 3.5	90.4 ± 10.9	92.3 ± 15.9
Language Skills (mean ± SD)	104.5 ± 7.9	95.5 ± 2.1	89.1 ± 10.8	92.5 ± 16.3
Motor Skills (mean ± SD)	89.5 ± 3.9	94 ± 12.8	80.4 ± 21	81.7 ± 13.2
Social-Emotional (mean ± SD)	98.3 ± 25.7	97.5 ± 3.5	100.5 ± 12.4	101.8 ± 17.6
Adaptive Behaviour (mean ± SD)	94 ± 17	102 ± 4.2	92.5 ± 10.8	86.3 ± 0

P1654 - SAFETY OF SILDENAFIL IN HEMODYNAMICALLY UNSTABLE CHILDREN

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Background: Sildenafil is a first-line agent used to treat pulmonary hypertension in pediatric patients. While it is often initiated in critically ill children, it is unknown whether sildenafil worsens hemodynamic instability in patients receiving continuous inotropic or vasopressor medications. We hypothesize that hemodynamic instability occurs frequently in children receiving concurrent vasoactive infusions after sildenafil initiation and that current sildenafil dosing recommendations may be inappropriate for this population.
Methods: Children younger than 2 years were included in this retrospective chart review if sildenafil was initiated during receipt of a vasoactive infusion between January 1, 2010 and September 30, 2016. Demographics, sildenafil dose, and vasoactive infusion data were collected. Patients receiving mechanical circulatory support or renal replacement therapy during the study period were excluded. The primary outcome was a composite endpoint of the incidence of sildenafil discontinuation and/or increased vasoactive support within 24 hours of sildenafil initiation.
Results: A total of 82 patients were included. The median age was 5 months (IQR 0.89-8.71) and 22% had a diagnosis of Trisomy 21. The median initial sildenafil dose for all patients was 1.29 mg/kg/day (IQR 0.86-2 mg/kg/day) with a median vasoactive inotrope score (VIS) of 5.28 (IQR 3.58-7.49) at sildenafil initiation. Twenty patients (24%) met the primary endpoint, 19 of which had inotropic support increased during the study period. Patients that met the primary endpoint tended to be younger (median 3.38 versus 5.5 months) with similar median total daily sildenafil dosing (1.13 mg/kg/day versus 1.33 mg/kg/day) as well as initial VIS (5 versus 5.5) between groups.
Conclusion: Critically ill children initiated on sildenafil while receiving concurrent inotropic infusions may be at risk for further hemodynamic instability. Lower empiric sildenafil dosing may be appropriate for this patient population to avoid the need for escalation of inotropic support.

P1680 - RISK FACTORS FOR MECHANICAL VENTILATION TIME AFTER CONGENITAL HEART SURGERY

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Background: Risk factors for prolonged mechanical ventilation time are not well defined and may lead to increased mortality, morbidity, and costs.
Objectives: To verify risk factors that affect mechanical ventilation time after congenital heart surgery by using the ASSSIST/PPSUSP databank of the state of São Paulo, Brazil.
Methods: Patients were prospectively included in the ASSIST/PPSUSP electronic databank, which gathers information from two tertiary hospitals where congenital heart disease surgeries. We retrospectively analyzed the data recorded from September 2014 to December 2015. We examined the following variables: weight

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at surgery, Prematurity, previous admission to intensive care unit (ICU), cardiopulmonary bypass (CPB) duration, procedure type, lactate levels after 12 hours at ICU, diagnosis of infection, and Rachs-1 classification. A Cox multiple regression model was used to correlate these risk factors and mechanical ventilation time.
Variables B p HR IC95%
Inferior Superior
Weight 0.014 0.000 1.014 1.009 1.018
Prematurity -0.156 0.403 0.856 0.593 1.234
Previous ICU -0.733 0.000 0.480 0.368 0.627
CPB duration -0.003 0.002 0.997 0.995 0.999
Procedure -0.399 0.173 0.671 0.378 1.192
ICU lactate -0.007 0.002 0.993 0.989 0.997
Infection -0.543 0.001 0.581 0.418 0.808
Rachs-1: 1-2 -0.241 0.104 0.786 0.588 1.050
Rachs-1: 3-6 -0.620 0.000 0.538 0.396 0.730
Conclusion: Previous ICU admission, CPB duration, UCI lactate, infection, and Rachs-1 higher than 3 affected mechanical ventilation time negatively in patients submitted to congenital heart disease surgery.

P1682 - PERINATAL ARRHYTHMIAS 10 YEARS EXPERIENCE

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Perinatal arrhythmias are frequent and usually benign. The aim of this study is to evaluate newborns with perinatal arrhythmia.
Material and Methods: Retrospective analysis of the clinical data of newborns admitted to neonatology due to cardiac arrhythmia, over a 10-years period.
Results: During study period, 113 newborns were admitted, 109 as primary arrhythmias and 4 as post-procedure complication. Of the 109 with primary arrhythmia, 62.4% were male, 12.8% (14) had congenital heart disease and 7.3% (8) other organs anomalies. Fetal diagnosis was made in 64.2% (70): 57.1% (40) tachycardia, 27.2% (19) premature contractions, 14.3% (10) bradycardia and 1.4% (1) tachy-brady syndrome. In complete AVB group, 7 mothers had autoimmune disease. Fetal hydrops was observed in 12. Prenatally drugs were prescribed in 22: 18 (45%)tachycardia (all digoxin;9 flecainide; 3 sotalol) and 4 (40%) bradycardia (4corticoid; 2salbutamol). Tachycardia termination was achieved in 11 on average 1.2 weeks after drug onset. Postnatal arrhythmia wasn´t observed in 10.
Thus, neonatal arrhythmia occurred in 99 (90.8%). Tachycardia in 65.6% (65) (28 AVRT – 11 WPW, 5 focal atrial tachycardia, 5 atrial flutter, 3 PJRT, 3 VT and 1 left fascicular), premature contractions in 22.3% (22) (15atrial and 6ventricular) and bradycardia in 12.1% (12) (10 cAVB, 1 2:1 AVB and 1 1° and 2° degree AVB). 12 (11%) had ventricular dysfunction. Within tachycardia group, anti-arrhythmic was given to 90.7% (59): 55 propranolol, 39 flecainide, 6 digoxin and 14 amiodarone iv. Electric cardioversion was performed in 10. Discharged antiarrhythmic therapy were maintained in 56. Follow-up time was 3 years and recurrences observed in 9 (13.8%). Within bradycardia group, 3 received isoprenaline, 6 temporary pacemaker and 7 definitive pacemaker. Mean follow-up time was 2.7 years (1 generator change and 1 remained without pacemaker). Seven (6%) children died, 3 related to arrhythmia.
Conclusion: Like other studies we conclude that perinatal arrhythmias are usually a a benign event,with low mortality and good long-term prognosis.

P1719 - IMPACT OF THE COMBINATION OF HIGH FLOW NASAL OXYGEN THERAPY AND INHALED NITRIC OXIDE ON THE POSTOPERATIVE MANAGEMENT OF THE FONTAN PROCEDURE

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Background: Since 2014, we have applied the combination of high-flow nasal oxygen therapy and inhaled nitric oxide after extubation to high-risk patients after the Fontan procedure. We investigated its benefits.
Materials and Methods: This was a single-center, retrospective review of 57 patients who underwent the Fontan procedure and needed nitric oxide inhalation therapy before extubation. Forty-nine patients were selected: patients who were ≧15 years old, or had preoperative tracheostomy, or postoperative brain complications, or postoperative phrenic paralysis were excluded. They were divided into two groups for comparison: early (January 2010 to December 2013, n = 28) and late (addition of high-flow nasal oxygen/ inhaled nitric oxide after extubation, January 2014 to June 2016, n = 21). There were no significant differences between the 2 groups in preoperative cardiac function and body weight, and they were similar for types of initial cardiac diagnosis: hypoplastic left heart syndrome (n = 6, 3), heterotaxy (n = 8, 11), and total anomalous pulmonary venous return (n = 6, 4). One patient in the late group had a single lung.
Results: There were no significant differences between the groups in intraoperative fluid balance, and central venous pressure at admission to the ICU. There were significant decreases in the late group in postoperative intubation time (53.1 ± 98.8, 5.9 ± 5.6 hours, p = 0.01), drainage time (24 ± 14, 15 ± 12 days, p = 0.01), and duration of the postoperative hospital stay (42 ± 22, 30 ± 16 days, p = 0.028), while urinary output per body weight at 6 hours post-surgery was significantly greater (p = 0.036). Three patients in the early group (10.3%) required re-intubation, but none was required in the late group.
Conclusions: Our results suggest that high-flow nasal oxygen therapy with inhaled nitric oxide contributes to a decreased requirement for re-intubation, and decreases the time required for postoperative intubation, drainage, and hospitalization.

P1729 - PREDICTION OF POST OPERATIVE INFECTION AFTER CONGENITAL CARDIAC SURGERY ROLE OF LAB PARAMETERS AND LOW CARDIAC OUTPUT STATE

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Aims and Objectives: To differentiate between infection and inflammation in post cardiac surgery. To predict the probability of infection in post operative Congenital cardiac condition based on clinical and lab parameters and low cardiac output (LCOS) state

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Methods: 210 patients who underwent cardiac surgery from October, 2015 till June, 2016 were enrolled in a prospective observational study. TLC, Platelet count, Procalcitonin, CXR, CRP done on day 1 and day 3 postoperatively along with cultures when indicated. Patients were classified into three groups. Patients with three or more abnormal values were included into group 3 (infection, n = 48), one or two criteria which normalized over next 48 hours into group 1(non infected, n = 111), all others into group 2(suspected infection, n = 51). Appropriate statistical analysis performed using SBSS software.
Results: Pre-operative respiratory infection and hospitalization were associated with post-operative infection. Post-operative CRP values failed to suggest infection. procalcitonin >2ng/ml and persistently higher levels on D3 was seen in group 3 as compared to group 2(p-0.000). Multiple regression analysis using TLC, fever, platelets, procalcitonin on D1, predicted infection with only 45% accuracy (F value- 27.22) and this went up to 70% accuracy(F value-45.23) if day 3 was included. LCOS on D1 is significantly more in group 3 as compared to group 2 (p- < 0.014).
Conclusion: It is difficult to differentiate between sepsis and inflammation in the post-operative period especially if cultures are negative. A combination of fever, chest x ray findings along with deranged TLC, platelet count, procalcitonin >2ng/ml can suggest infection with an accuracy of 70% on D3 as suggested by regression analysis. LCOS on D1 is possibly the best predictor of possible infection. We believe that having covered BSI, VAP, and CAUTI, with appropriate strategy, the only other source of infection is transmigration of bacteria from relatively ischemic gut exacerbated by the LCOS.

P1743 - INCESSANT JUNCTIONAL ECTOPIC TACHYCARDIA AS INITIAL PRESENTATION OF ENTEROVIRAL MYOCARDITIS SUCCESSFUL BRIDGING TO RECOVERY WITH ECMO SUPPORT

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Junctional Ectopic tachycardia (JET) is a rare presentation in neonatal myocarditis. We report a case of neonatal enteroviral myocarditis and encephalitis, who presented with fever and JET, and refractory to direct-current cardioversion and antiarrhythmic therapy, including amiodarone, esmolol and ivabradine. Subsequently the clinical course progressed to ventricular tachyarrhythmia and severe ventricular dysfunction, requiring emergency support using extracorporeal membrane oxygenation (ECMO). Intravenous immunoglobulin was given but resulted in significant platelet aggregation and fibrin formation, complicated with haemolysis and haemoglobinuria requiring exchange of circuit. Patient recovered after 15 days of ECMO support with full recovery of cardiac function and resumption of sinus rhythm, with satisfactory neurological outcome. A literature review of 16 cases of neonatal enterovirus myocarditis from year 2008-2016 was performed, showing a mortality of 31%, similar to previous literature review. JET is a rare presentation even in cases presented with arrhythmias. Role of IVIG remains controversial in viral myocarditis and risk of thromboembolic events has to be considered. ECMO support and heart transplantation are the ultimate management for neonatal viral myocarditis refractory to medical therapy.

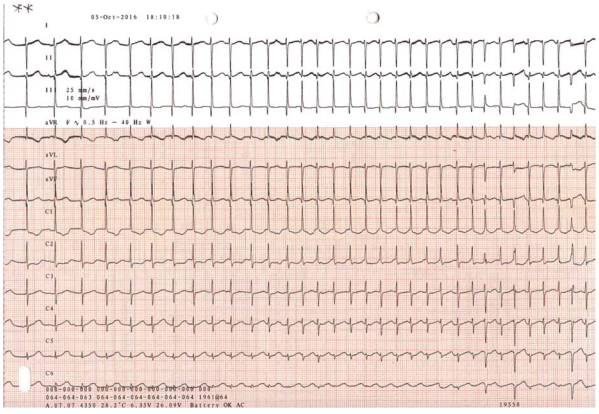

Figure 1.

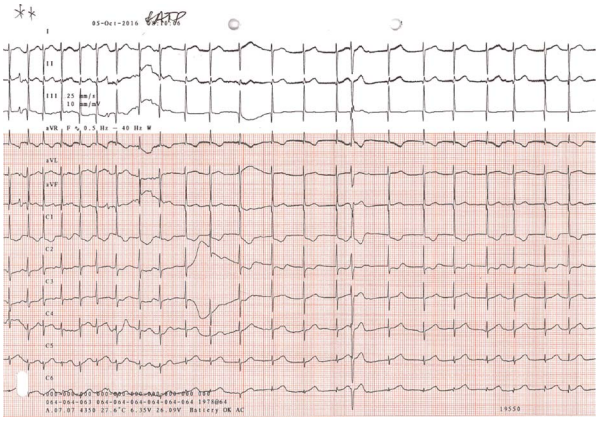

Figure 2.

P1745 - INCIDENCE AND RISK FACTORS FOR EARLY POST OPERATIVE ARRHYTHMIAS FOLLOWING PEDIATRIC CARDIAC SURGERY

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Background: This prospective study was conducted to determine the age specific incidence and the risk factors for early post operative arrhythmias in the intensive care unit following pediatric cardiac surgery with special focus on the disease specific incidence and early outcome using a uniform strategy for management of the arrhythmias and a special reference to serum magnesium levels in the post operative period.
Methods: The study population included 370 consecutive pediatric patients undergoing open heart surgery using cardiopulmonary bypass, from September 2013 to July 2015 regardless of the diagnosis. Patients having pre operative arrhythmias were excluded from the study. Patients were continuously monitored, and hemodynamically significant arrhythmias were recorded.
Results: The incidence of post operative arrhythmias was 11.12% (n = 25). The predominant arrhythmias noted were junctional ectopic tachycardia (JET) (n = 15, 60%), ventricular premature ectopics/ ventricular tachycardia (n = 8, 32%), supraventricular

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tachycardia (n = 2, 8%), ventricular fibrillation (n = 2, 8%) and atrial fibrillation (n = 1, 4%). 22 patients recovered and were discharged uneventfully and 3 deaths were reported in the study group. Factors that were found to be significantly related statistically to the occurrence of arrhythmias included surgical duration (p = 0.01), serum lactate (p = 0.04), systolic blood pressure (p = 0.02) and Wernowsky inotropic score (p = 0.02). Serum electrolyte levels (sodium, potassium, magnesium and calcium) were not significantly different in the two groups. Both atrioventricular septal defect repairs (n = 4) and arterial switch operations (n = 4) had equal incidence of the arrhythmias in the post operative period.
Conclusions: We found a low incidence of arrthymias particularly JET after pediatric open heart surgery. A management protocol consisting of amiodarone, digoxin, surface cooling and optimization of inotropes was uniformly used and found to be effective. Longer surgical duration, higher serum lactate levels, lower systolic blood pressure and higher Wernowsky inotropic score were found to be independent risk factors for the early post operative arrhythmias.

P1746 - NOSOCOMIAL INFECTION RELEVANCE AND OUTCOME IN INFANTS UNDERGOING OPEN HEART SURGERY IN THE PRESENT ERA

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Background: To evaluate the causal relation between nosocomial infection(NI) and clinical outcomes following cardiac surgery in neonates & infants and to identify risk factors for the development of NI in this subset of patients
Methods: After Ethics committee approval, one hundred consecutive infants undergoing open heart surgery (OHS) between June 2015 and June 2016 were included in this prospective observational study. Data were prospectively collected using a standardized data collection form. The incidence and distribution of NI, the microorganisms, their antibiotic resistance and patient outcome were determined. Centers for Disease Control and Prevention criteria were used for defining NIs.
Results: Sixteen infants (16%) developed microbiologically documented NI after cardiac surgery. Neonatal age group was found to be most susceptible. Lower respiratory tract infections accounted for majority of the infections (47.4%) followed by bloodstream infection (31.6%), urinary tract infection (10.5%) and surgical site infection (10.5%). Klebsiella (36.8%) and Acinetobacter (26.3%) were the most frequently isolated pathogens. Major risk factors for NI were neonatal age group ([odds ratio] OR = 6.4), preoperative hospital stay ≥3 days (OR = 3.7), duration of cardiopulmonary bypass >180 minutes (OR = 5), aortic cross clamp >99 minutes (OR = 2.8), open sternum ≥2 days(OR = 4.4) and surgical complexity score (P = 0.03).
Conclusion: NI still remains a dreaded complication in infants after OHS and contributing to morbidity and mortality. Strategies like decreasing preoperative hospital stay, early sternal closure, early extubation and rational antibiotic usage should be implemented to prevent NI.

P1781 - IS PRE OPERATIVE ENTERAL FEEDING SAFE IN INFANTS WITH DUCT DEPENDENT CONGENITAL HEART LESIONS

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Background: Infants with duct-dependent congenital heart lesions are treated pre-operatively with a continuous prostaglandin E1 infusion to maintain patency of the ductus arteriosus. The optimal feeding strategy of these patients remains controversial, and there is widespread variation in practice worldwide. Many units choose not to feed these patients enterally whilst on a prostaglandin infusion due to the theoretically increased risk of necrotising enterocolitis (NEC), although there is a lack of high-quality evidence to demonstrate this association. We aimed to describe the feeding strategies used at the Royal Children's Hospital in infants receiving prostaglandin, and to describe the incidence of gastrointestinal complications (including NEC) in this patient group, investigating whether enteral feeding is associated with a higher risk.
Methods: Using an institutional cardiology database, all patients diagnosed with hypoplastic left heart syndrome, coarctation of the aorta, pulmonary atresia, or transposition of the great arteries born between September 2013 and December 2014 were identified. Data were gathered from the hospital records. Premature infants, those with pre-existing gastrointestinal disease, and those who never received prostaglandin were excluded. Data were compared using the chi-squared, Fisher's exact, and Student's T test as appropriate.
Results: 89 infants were identified (11 hypoplastic left heart, 28 coarctation, 14 pulmonary atresia, 36 transposition). 7 patients received a clinical diagnosis of suspected or confirmed NEC (incidence 7.9%). 26 (29.2%) patients received some enteral feed whilst receiving prostaglandin. There was no association between the diagnosis of NEC and enteral feeding (P = 0.1), sex (P = 0.1), timing of diagnosis (P = 0.9), cardiac diagnosis (P = 0.3), or birthweight (P = 0.6). All NEC cases were managed conservatively.
Conclusions: Based on these retrospective non-randomised data, there does not appear to be an association between enteral feeding and NEC in infants receiving prostaglandin. Further prospective studies are required to confirm the safety of enteral feeding in this patient group.

P1791 - DILATED CARDIOMYOPATHY DUE TO SEVERE IRON DEFICIENCY ANEMIA

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Background: Alimentary iron-deficiency is the most common cause of nutritional anemia in both developed and developing countries. Typical symptoms are fatigue, exercise limitation and tachycardia. We present a case of dilated cardiomyopathy due to severe iron deficiency anemia.
Results: A previously healthy 23 months old girl came to our emergency department with a recent history of peripheral edema, fatigue and dyspnea. During the last 12 Months she was fed exclusively with 1.5% cow's milk and fruit juice. Physical examination revealed hepatomegaly, pallor and a systolic murmur with

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hyperdynamic precordium. Lab results showed a low hemoglobin level of 1.4 g/dl and a hematocrit level of 7.1%, the mean corpuscular volume was 66fl (normal range 73-101fl), mean corpuscular hemoglobin was 13 pg (normal range 23-31 pg). Ferritin was below detection limit, Vitamin B12, B1 and folic acid were within normal range. There were no signs of bleeding, hemolysis or inflammation. Initial NT-proBNP was 14258 pg/ml (normal range <320 pg/ml). Echocardiography revealed severe dilation of the left ventricle (LVIDd 46mm, Z-Score +4.4) and mild to moderate LV dysfunction with fractional shortening of 24% and ejection fraction of 48%, mild mitral regurgitation and a tricuspid regurgitation peak gradient of 40 mmHg. The patient received 2 units of packed red blood cells and was treated with oral iron sulfate, an initial short course of Furosemide followed by Lisinopril and Bisoprolol. Within 12 days cardiac function and LV-diameter improved markedly (LVIDd 38mm, Z-Score +2.5, fractional shortening 31%, ejection fraction 59%, peak tricuspid regurgitation gradient 25 mmHg).
Conclusion: Iron deficiency is the most common cause of nutritional anemia. A strict vegetarian/vegan lifestyle can be a contributing factor. The exact pathomechanism of anemia induced cardiomyopathy remains unclear, but might be explained by tissue hypoxia and chronic increased sympathetic nervous activity. Fortunately, blood transfusion, iron substitution and anticongestive medication usually result in complete restitution.

P1803 - CARDIAC THROMBI IN NEWBORN TREATMENT WITH RECOMBINANT TISSUE PLASMINOGEN ACTIVATOR TEN YEAR EXPERIENCE

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The incidence of cardiac thrombi in newborns has increased with the use of central venous catheters. Preterm infants have high risk of catheter-related thrombus formation due to a combination of high prothrombotic activity, low levels of natural anticoagulants and an imbalance of the fibrinolytic system. Thrombolysis with recombinant tissue plasminogen activator (rTPA) has been used as an alternative to heparin in life threatening thrombus with risk of embolization. The objective was to study the response and complications of rTPA in the management of life threatening cardiac thrombi in newborns.
Materials and Methods: 10 year retrospective study, 11 medical records of newborns with intracardiac thrombi were analyzed.
Results: 10 patients were premature newborns, median age was 30 weeks, median weight was 1003 g (ranging 856-3430 g) and 6 weighed less than 1000 g. Diagnosis age was between 5 to 18 days. 10 subjects suffered from sepsis and all of them had central venous catheters. 10 thrombi were located in the right atrium and one in the left atrium. Three patients received low molecular weight heparin prior rTPA. 5 patients showed complete resolution of thrombus, 4 partial resolution and 2 did not respond to therapy. One of these last required thrombectomy and the other died from sepsis. Four patients had intracranial hemorrhage grade I without sequelae.
Conclusion: The use of rTPA is an effective therapy in newborn afflicted with cardiac thrombi. This therapy achieved the resolution or reduction of the thrombus and therefore, reduced the secondary life-threatening risk without serious side effects. The thrombolysis was effective even in newborn weighing less than 1000 g. New prospective studies are required to further validate the use of rTPA.

P1827 - PATIENT DEMOGRAPHICS AND EARLY OUTCOMES AFTER CARDIOPULMONARY RESUSCITATION IN PAEDIATRIC CARDIAC INTENSIVE CARE UNIT

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Background: Children with underlying cardiac problems are at greater risk of experiencing cardiac arrest compared to general population with reported incidence of 2-6%.
Objective: Describe patient demographics, aetiology, and early outcomes after cardiopulmonary resuscitation (CPR) in a tertiary-care, Paediatric Cardiac Intensive Care Unit (PCICU).
Methods: A retrospective, cohort study of children (<16 years) admitted to the PCICU who underwent CPR between 31/5/13–12/31/15. Outcomes assessed were return of spontaneous circulation (ROSC), ECMO cannulation during CPR (ECPR), and in-hospital mortality.
Main Results: There were 39 CPR events in 2145 children (1.8%) during the study period. Patient demographics are given in Table 1. Successful resuscitation (ROSC or ECPR) was possible in 79.5% (ROSC-64.1% and ECPR-15.4%). In-hospital survival

Table.

Patient Demographics

Age	1.5 months (4 day-15 year)
Sex (Female)	24/39 (61.5%)
Weight (kg)	3.2 (2-88)
Unplanned Admission	20/39 (51.3%)
Night Shift (8pm-8am)	24/39 (61.5%)
Not Ventilated	5/39 (12.8%)
Difficult airway	3/39 (7.7%)
Surgery	24/39 (61.5%)
Risk adjusted congenital heart surgery score (RACHS) Category (n = 22)	
2	2 (9.1%)
3	11 (50.0%)
4	5 (22.7%)
6	4 (18.2%)
Systemic to PA shunt	10/39 (25.6%)
Preceding rhythm	
Bradycardia	30/39 (76.9%)
Ventricular Fibrillation/Tachycardia	6/39 (15.4%)
Paced-Complete Heart Block	2/39 (5.1%)
Junctional Ectopic Tachycardia	1/39 (2.6%)
Vasoactive Inotrope Score prior to event	8 (0-70)
Lactate prior to event (mmol/L)	7 (0.3-18)
Acute Kidney Injury- preceding	13/39 (33.3%)
Inhaled Nitric Oxide	9/39 (23.1%)

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was 61.1% overall: 82% with ROSC and 50% requiring ECPR survived to hospital discharge. Survival was better in surgical compared to medical admissions (73.9% vs. 38.5%, p = 0.036). Approximately half of the CPR events occurred within 24 hours of PCICU admission. Comparing survivors to non-survivors, the duration of CPR (5.5 (0-45) minutes vs. 11(0-53) minutes, p = 0.15) and the occurrence of CPR at night (8 pm-8 am) (59.1% vs. 40.9%, p = 0.75) were not significantly different, respectively. Higher serum lactate and vasoactive-inotrope score (VIS) prior to CPR was associated with mortality. In surgical patients with CHD, all in-hospital deaths occurred in children with a systemic to PA (SPA) shunt.
Conclusion: We were able to identify patient factors in our unit associated with CPR events and poor outcomes: SPA shunts, higher VIS and lactate levels prior to CPR, and preceding end-organ dysfunction. Critical time periods: 24-48 hours post- surgery and after delayed sternal closure were also identified. A multi-disciplinary approach is being implemented to prevent need for CPR and improve outcomes after CPR based on these findings.

P1834 - THROMBOELASTOGRAPHY CAN PREDICT RISK OF BLEEDING IN PATIENTS ON EXTRACORPOREAL MEMBRANE OXYGENATION SUPPORT

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Background: Patients undergoing Extracorporeal Membrane Oxygenation (ECMO) are at high risk for complications of bleeding and thrombosis. Current methods of laboratory assessment of the coagulation system (PT/PTT, ACT) are imprecise and thereby complicate clinical decision-making. We hypothesize that assessment of physical properties of clot formation by thromboelastography (TEG) is more predictive of bleeding than PTT for patients on ECMO. In addition, thrombin –antithrombin (TAT) -complex, fibrinogen and plasminogen function coagulation tests maybe be useful in predicting bleeding in patients on ECMO.
Methods: In a prospective study at a single institution, TEG was performed in patients with congenital heart disease requiring ECMO support. Whole blood samples were collected 6-12hrs following ECMO initiation and daily for the duration of support. Platelet poor plasma was utilized for the functional and immunoassays. Bleeding during each 24hr period was defined as need for re-exploration and transfusion requirement. Associations between TEG parameters and bleeding over each 24hr period (bleeding vs. non-bleeding days) were assessed using logistic regression analysis.
Results: Twenty patients were enrolled, yielding a total of 116 patient-days of data for analysis. TEG MA, but not PTT, was associated with bleeding (p = 0.03 and p = 0.1 respectively). Fibrinogen levels were significantly lower in patients on bleeding days compared to non-bleeding days (320 mg/dl (IQR: 229-412) vs. 376 mg/dl (IQR: 306-510), p = 0.02). Abnormally elevated plasminogen function (>100%) and TAT levels (>10 micrograms/ml) were more frequently observed on bleeding days compared to non-bleeding days in patients on ECMO (Table 1).
Conclusions: In patients undergoing ECMO support, TEG MA and fibrinogen levels correlate better with bleeding than does PTT. Elevated TAT and plasminogen function suggests that hyperactive thrombin generation and fibrinolysis are present on bleeding days compared to non-bleeding days. TEG may be a useful diagnostic test in monitoring coagulation system in these patients.

Table 1. Abnormal coagulation markers comparing bleeding days verses Non-bleeding days in patients on ECMO.

Coagulation Markers	% Bleeding days	% Non-Bleeding days	p-value
Low TEG MA (<50 mm)	83	50	0.03*
Elevated PTT (>60 seconds)	65	47	0.1
Elevated Plasminogen Function (>100%)	27	10	0.04*
Elevated TAT- complex levels (>10 μg/ml)	86	65	0.02*

*p-value < 0.05 is considered significant.

P1869 - COARCTATION OF THE AORTA IN NEWBORN

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Background: Coarctation of the aorta represents 5-7% of all congenital heart diseases. Early, at best, prenatal diagnosis permits to improve prognosis. The objective of this study is to analyze epidemiological, clinical, therapeutic aspects as well as outcome of the constrictions of the aorta in neonatal period.
Methods: We conducted a descriptive study, enrolling all the patients with constriction of the aorta diagnosed and confirmed in prenatal or neonatal period, over 12 years, in the department of neonataology of Farhat Hached teaching hospital (Sousse).
Results: Twenty seven newborns had a constriction of the aorta representing 6.47% of congenital cardiac malformations with an incidence of 0.16% alive births. Diagnosis was suspected in prenatal period in only one case. Clinical features suggestive of a congenital heart disease were mainly circulatory insufficiency features or respiratory symptoms not explained by a pulmonary cause. Cardiac ultrasound allowed making the diagnosis of the constriction in 23 newborns. The preoperative mortality rate was of 35% and was in relation with the cardiac malformation in 71.4% of cases. Twenty neonates survived and underwent surgery. Post operative mortality rate was of 35% with a total mortality of 51.8%. The rate of coarctation relapse among survivors was 30.8%.
Conclusion: Prognosis of the constrictions of the aorta in neonatal period remains reserved in our department. It could be improved thanks to the progress of diagnostic and therapeutic means.

P1872 - PULSE OXIMETRY SCREENING FOR CRITICAL CONGENITAL HEART DISEASE IN NEWBORNS

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Introduction: Congenital heart disease (CHD) are the commonest type of congenital malformation, occurring in 6– 8 per 1000 live births. 25% of them are Critical (CCHD), needing surgery or other procedures in the first year of life. Screening is important; it provides significant opportunity to reduce infant mortality and morbidity. Considering that routine clinical examination and antenatal ultrasound still miss a significant proportion of cases. Based on the rationale that most newborn babies with CCHD have a degree of hypoxemia, the use of pulse oximetry to screen

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asymptomatic babies for CCHD is Simple, painless, non-invasive. The aims of our study were to determine the CCHD's incidence, screened by pulse oximetry added to physical assessment and to compare it with the incidence evaluated before the introduction of this technique in the screening protocol.
Methods: Prospective screening study of all babies born in Susah's maternity hospital during 12 month (1er February 2014-31 January 2015).
Results: During the time of this study a total of 7992 live born were enrolled to the study. The incidence of CCHD were 3.25‰ (n = 26). This incidence is greater than that detected by the clinical assessment alone (p = 0.025). Screening was performed by pulse oximetry in 5 infant and by routine clinical examination in 21. Two infants have been positive POX screening without CCHD. They had pulmonary infection, so the specificity of this technique was 99.99% with positive predictive value 71.4%.
Conclusion: Pulse-oximetry screening offers an effective, accurate and reliable means for detecting CCHD in asymptomatic newborns. It should be used, added to clinical examination, in maternity hospitals as a screening method. Next step: multicenter study.

P1873 - SHORT AND LONG TERM OUTCOMES OF PRIMARY NEONATAL REPAIR FOR TETRALOGY OF FALLOT PULMONARY STENOSIS VS. PULMONARY ATRESIA

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Background: The management of symptomatic neonates with tetralogy of Fallot and pulmonary stenosis (TOF/PS) or pulmonary atresia (TOF/PA) and well-developed native pulmonary arteries remains controversial. Since 1997, our institutional approach was to perform primary neonatal repair if the preoperative weight was over 2.5 kg. Right ventricular outflow tract (RVOT) was reconstructed without use of valved conduit except for cases of unfavourable coronary anatomy and complete absence of pulmonary artery trunk. We sought to determine short- and long-term results of this strategy and compared outcomes of TOF/PS and TOF/PA subgroups.
Methods: This is a retrospective review of 35 neonates undergoing primary repair at a single centre (1997-2016). Fourteen TOF/PS and 14 TOF/PA patients underwent transannular/transjunctional patch repair and 4 TOF/PS patients received valve-sparing RVOT reconstruction. In 2 neonates with TOF/PA and 1 TOF/PS, the primary conduit placement was needed.
Results: The median age at primary repair was 16 days for TOF/PS and 8.5 days for TOF/PA (p = 0.02). One patient with TOF/PS died early after surgery. Postoperative course was comparable for TOF/PS and TOF/PA subgroups according to duration of mechanical ventilation (96 vs. 111 hours; p = 0.86), maximum vasoactive-inotropic score (10 vs. 10.5; p = 0.27), incidence of junctional ectopic tachycardia (33.3% vs. 18.7%, p = 0.33) and hospital stay (19 vs.15 days; p = 0.72). One infant with TOF/PA died during follow-up. One third (33%) of patients underwent RVOT/pulmonary arteries reintervention during mean follow-up of 8.8 + /-6.2 years. Overall freedom from RVOT reintervention was 88.9% in TOF/PS and 64.3% in TOF/PA at 5 years and 79.0% in TOF/PS and 57.1% in TOF/PA at 10 years (p = 0.17).
Conclusions: Primary repair of TOF/PS and TOF/PA with present native pulmonary arteries can be safely accomplished without use of conduit placement in majority of normal weight neonates. Late reintervention rate is reasonable and tends to be higher for TOF/PA group.

P1876 - DUCTUS ARTERIOSUS IN PRETERM INFANTS DO WE REALLY NEED TO TREAT IT

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Introduction: There is still controversy about the optimal treatment in management of the ductus arteriosus of the preterm infants.
Objective: to analyze the results of ductal closure in extreme preterm infants in a Neonatal Unit in a developing country.
Patients and Methods: Between December 2015 and December 2016, we had 104 patients younger than 32 weeks. All had an echocardiogram at 48-72 hrs. of life DAP > 3 mm, AI/Ao > 1.5, reverse diastolic flow in the descending Ao, estimated GC of VI > 300 ml/Kg / min.
Results: Of the 104 patients, only 48 had a PDA that fulfilled these criteria (46%). Of these 22 received pharmacological treatment (21%): 10 were treated with paracetamol (mean gestational age 26.6 wk, 24-29), 1 with ibuprofen (24 wk) and 11 with indomethacin (mean gestational age: 27.3 wk, 23-31), observing its closure by 90% cases. Only 2 patients of this group underwent surgical ductal closure (1,9%). The other 26 patients were followed until their spontaneous closure, considering that they did not use NSAIDs because there was an intercurrent infection, excess fluid supply or inadequate mechanical ventilation.
Conclusion: currently only a small percentage of patients require ductal pharmacological closure,even at extreme gestational ages, when they are individually selected and with specific echocardiographic criteria, reducing their cost of treatment and the potential complications of NSAID use.

P1887 - FREQUENCY AND OUTCOME OF ACUTE NEUROLOGIC COMPLICATIONS IN PATIENTS AFTER CONGENITAL HEART DISEASE SURGERY

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Objectives: To determine the frequency and immediate outcome of acute neurologic complications in children undergoing congenital heart surgery
Methods: This retrospective unmatched case control study was done at our 4 bedded pediatric cardiac intensive care unit. All patients from newborn upto 30 years who underwent congenital heart surgery from January 2010 to October 2016 and developed a neurologic complication (seizure, abnormal movements, altered sensorium, stroke) were included. Data collected included cardiac diagnosis, anatomical lesion, type of surgery, cardiopulmonary bypass time and aortic cross clamp time, GCS, neurophysiologic and radiologic investigations done and outcome. Data was entered and analyzed using SPSS v 20. Chi square test was used to determine the association of different factors associated with neurological problems. P value of <0.05 was taken as significant
Results: A total 35 patient's developed neurological complication during the study period (35/1300, 2.7%) for which 35 controls were chosen based on similar time of admission. Both the group were similar in age (Median age 18 and 21 months for cases and

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controls), gender (males were 25 and 22 in cases and controls), cyanotic congenital heart disease (n = 23 and 24 in cases and controls), 12 had single ventricle physiology, and 29 patients surgery was done on cardiopulmonary bypass machine in cases and 30 in controls. 28 patients developed seizures postoperatively, 5 had altered sensorium and 2 patients developed stroke while 6 of these developed brain death and expired and 5 others had poor Glasgow coma outcome scale. Cardiopulmonary resuscitation, prolonged aortic cross clamp time were significantly ($p < 0.05$) associated with development of neurological complications.
Conclusion: Neurological morbidities after congenital heart surgery are uncommon but significant contributor to morbidity of these patients.

P1891 - SYSTEMATIC REVIEW AND META ANALYSIS ON MANAGEMENT OF PEDIATRIC STROKE

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Although acute ischemic stroke (AIS) in children is rare, the disease poses a significant clinical problem due to its potentially devastating consequences and its complex pathophysiology. While treatment in adults is well established, evidence in children is limited to date. We reviewed the current literature and synthesized data on safety and efficacy of intravenous thrombolysis (IVT) and endovascular therapy (EVT) in pediatric AIS. We performed a systematic review and meta-analysis of all available case series and observational studies that evaluated safety of IVT and EVT in stroke patients aged <18 years, since there were not enough papers to be included only with children with congenital heart disease. We searched Medline/PubMed, Cochrane Library, Google Scholar for eligible studies. Safety outcomes included any intracerebral hemorrhage post-treatment and in-hospital mortality. A random-effects model was used to compute pooled effect estimates and the I2 statistic was used to assess heterogeneity. Our analysis complied with PRISMA statement. We identified 152 records, of which only 3 studies with a total of 16,335 pediatric patients with AIS met our eligibility criteria. Of these studies, two explored safety of sole IVT and one combinatory IVT/EVT. In-hospital mortality rates were similar between pediatric stroke patients treated with either IVT or IVT/EVT and controls (odds ratio = 0.85, 95% CI: 0.15-4.87; $p = 0.857$), with moderate evidence of heterogeneity (I2 = 64%). Risk of intracerebral hemorrhage was substantially increased in IVT (odds ratio = 3.60, 95% CI: 1.66-7.80; $p = 0.001$) compared with controls, with no evidence of heterogeneity (I2 = 0%). Efficacy of revascularization therapies could not be analyzed due to lack of uniform outcome data in the included studies. We revealed a substantial lack of evidence for acute revascularization treatment of pediatric AIS. While an increased risk of intracerebral hemorrhage related to IVT emerged in our analysis, further research is needed to elaborate these findings and identify the optimal treatment regimen for pediatric stroke.

P1905 - ECMO IN POST HEART SURGERY PEDIATRIC PATIENTS.A CASE REPORT.CHRISTUS MUGUERZA HOSPITAL MONTERREY N.L

María Verónica Contreras Cepeda, Rene Gómez Gutiérrez, Gerardo Vargas Camacho, Francisco González, Guillermo Quezada Valenzuela
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Introduction: Extracorporeal Membrane Oxigenation (ECMO) is a life support procedure where cardiopulmonary function is substituted by a machine. ECMO is indicated in case of serious respiratory insufficiency, or cardio respiratory insufficiency without response to current treatment. Since 1980's, ECMO has been a therapeutic alternative in third level hospitals in America and Europe, but it hasn't been used in Mexico. In 2013 Neonatal ECMO Program was initiated in Christus Muguerza Hospital in Monterrey Mexico. Results are described.
Materials and Method: This is an statistical, retrospective, observational, and analytical study. ICU Patient's clinical files in Christus Muguerza Hospital Monterrey were analyzed since January 2013 up to date. Cardiac surgery immediate post operatory patients under ECMO were included. Patient's incomplete files were excluded. Data were collected in an Excel Database and were analyzed using SPSS Statistics Program.
Results: Most of the patients who entered into ICU were newborn (54.5%); most of them were male. The ICU admission diagnose was low cardiac load, with or without lung failure. The most common causes of cardiopathy were Great Vessels Transposition (GVT) and Tetralogy of Fallot (TOF). Co morbidity before ECMO was variable, and Dimorphic Syndromes were frequent, except in 3 patients who didn't have any co morbidity. The most common complications were acidosis, bleeding and infection. Respiratory acidosis was the most frequent. Pneumonia and sepsis were the most frequent type of infections. The current treatment was integrated by antibiotics (Meropenem), amines (Milrinone), anticoagulants (Heparine), and blood products.

P1906 - ECMO POSTCARDIOTOMY IN A NEWBORN WITH COMPLEX CONGENITAL CARDIAC DISEASE FIRST SUCCESSFUL CASE IN MEXICO AND NEUROLOGICAL OUTCOME AT 18 MONTHS

María Verónica Contreras Cepeda[1], Adriana Nieto Sanjuanero[1], Bárbara Cardenas Del Castillo[1], Rene Gómez Gutiérrez[1], Guillermo Quezada Valenzuela[1], Victor Quezada Elvira[1], Adrián Abrego Ambía[1], Arturo Saldivar[1], Jaime García[2], Teresa Medina[2]
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Introduction: Extracorporeal Membrane Oxygenation (ECMO) is a life support therapy that temporarily replaces cardiopulmonary function.
Clinical Case: Male neonate, 37 weeks, 2770 gr, Apgar 8-8, cyanosis at birth. Echocardiogram/Angio-CT: Taussig-Bing anomaly with Severe Aortic Arch Hypoplasia, large PCA and LV outflow tract obstruction by myxomatous mass in the mitral valve. On the 7th day of life, they performed corrective surgery, with a score of 4 in RACHS-1 Scale, performing aortic arch repair, IV closure, Jatenne type arterial swich, myxoma resection and IA closure with extracorporeal circulation time (CEC) 392 Minutes, Aortic Time-Clamping 198 minutes, Selective Time-Brain Perfusion 24 minutes, Temperature 25oC. In the attempt to exit extracorporeal circulation (ECC) presented complete AV block and severe ventricular dysfunction, started ECMO VA support with central cannulation providing ECMO pump flow 100 ml/kg/hour with immediate recovery of blood pressure and systemic perfusion. The thorax was covered with plastic and transported in ECMO to the NICU. Twenty-four hours post-ECMO-VA , the patient regain pulsatility and sinus rhythm with normal lactate values. Anticoagulation with UF heparin to maintain anti Xa levels 0.2 to 0.5 UI/mL., At 3rd day on ECMO support with 60% LVEF, weaning from ECMO-VA was successfully done. Hospital stay 22 days, follow-up by multidisciplinary team without complications after

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discharge. At 18 months a Bayley III Development Scale was made with a score by areas: Cognitive 95, Language 106 and Motor 107.
Discussion: ECMO-VA is a valuable therapy in postoperative patients with complex congenital heart disease with prolonged CPB and failure to exit ECC.
Conclusions: ECMO programs in Congenital Heart Surgery Centers are necessary in Mexico , they increase survival. It is important to perform long-term neurological follow-up.

P1922 - USING SOCIAL MEDIA TO OBTAIN PARENTAL FEEDBACK ON COMPLICATIONS ASSOCIATED WITH PAEDIATRIC CARDIAC SURGERY A QUALITATIVE ANALYSIS OF DATA FROM A CHARITY MODERATED ONLINE FORUM

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Background: As mortality rates fall it becomes increasingly important to focus on morbidity associated with paediatric cardiac surgery, including how information about morbidity is communicated to parents. Our aim was to elicit parents' understanding and experience of complications following their child's cardiac surgery, using an online forum as a means of accessing viewpoints that may be missed in alternative data collection methods.
Methods: The Children's Heart Foundation (a national parent charity accessed by families at all UK paediatric cardiac centres) set up and moderated a closed, anonymous online discussion group via their Facebook page, which focused on complications, information needs and methods of providing families with information. The forum transcript was thematically analysed by the research team.
Results: The forum ran over 3 months and involved 72 participants (68 mothers, 1 father, 1 patient, 2 grandmothers; age range 15-59 years). Several themes were identified:

- Language used by clinicians about complications of surgery may be poorly understood by families.
- There can be a lack of consistency in how we explain complications.
- Parents often felt unprepared for complications when they arose.
- There is no 'right amount of information' that suits every family.
- Written and audio-visual material about surgery and expected complications, as well as access to a liaison staff member, would be helpful aids.

Conclusions: Our language is often misunderstood. There is wide variability in the way we describe complications, much of which is not absorbed or retained. Adjuncts and availability of a liaison staff member may help understanding and preparation for surgery, intensive care and beyond. The nature of the online forum promoted free debate and sharing of views, providing rich data that may not have been elicited in face to face groups or interviews. These data have helped us identify and understand areas for improvement.

P1926 - ASSOCIATION BETWEEN RACE AND ILLNESS SEVERITY IN CHILDREN UNDERGOING CARDIAC SURGERY

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Background/Hypothesis: Prior studies have demonstrated that non-white race/ethnicity and insurance status are associated with mortality after congenital heart surgery. We hypothesize that non-white children undergoing congenital heart surgery have higher severity of illness and, in turn, have higher mortality and ICU lengths of stay than their white counterparts.
Materials and Methods: We performed a retrospective analysis of registry data from the Virtual Pediatric Systems dataset from 2009-2016. All children (less than 18 years of age) undergoing cardiac surgery and admitted to an ICU in one of the 82 participating institutions were included (n = 30, 814).
Results: In a univariate analysis, there was significant variation in severity of illness scores across race/ethnicity groups with black, Hispanic and other race/ethnicity patients having higher severity of illness scores than white patients. However, multivariate regression models did not demonstrate an association between race/ethnicity and severity of illness scores, after adjustment for insurance status and other covariates. In multivariate models that examined ICU mortality, black patients remained at increased odds of mortality (Odds Ratio: 1.38, 95% Confidence Interval: 1.09-1.75) when compared to white children, adjusting for illness severity, insurance, gender, complexity of cardiac surgery, weight, and age. In a multivariate regression model for ICU length of stay, black children (Estimate: 0.87, 95% Confidence Interval: 0.08-1.65) and children on government insurance (Estimate: 1.30, 95% Confidence Interval: 0.34-2.25) experienced longer lengths of stay when compared to white patients and patients with private insurance.
Conclusions: Black children undergoing cardiac surgery have higher odds of death compared to white children, despite adjustment for illness severity on admission to the ICU. These results suggest that severity of illness may not be the main driver of health disparities in children undergoing congenital heart surgery.

P1989 - LUNG ULTRASOUND (LUS) TO ASSESS PERSISTENT PULMONARY HYPERTENSION OF THE NEWBORN (PPHN) ETIOLOGY

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Background: PPHN is defined by the failure of post-natal reduction in pulmonary vascular resistance with significant morbidity and mortality associated. Patients may require Extracorporeal Membrane Oxygenation therapy (ECMO) although not having a causal diagnosis. LUS has proved to be extremely useful to diagnose pulmonary diseases. The aim of this study was to explore LUS pattern in newborns with PPHN requiring ECMO. We hypothesized that patients with a normal LUS had a non lung parenchimal disease and that LUS was as useful as CXR to reach the causal diagnosis.
Materials and Methods: This is a prospective study of LUS to assess PPHN etiology in patients admitted for ECMO, with patients recruited from 2014 to 2016 at Hospital Sant Joan de Déu at the NICU. LUS was performed by a neonatologist blinded to the perinatal history before the patient was placed in ECMO, scanning four lung areas in each hemithorax;

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afterwards they were compared with CXR diagnosis. PPHN final diagnosis was made based on the patient's medical history, and complementary tests, without the information provided by LUS.
Results: Fifteen patients were recruited. Ten were male (66.7%) and the median gestational age was 38.3 weeks, with eleven term newborns (80%). Twelve were under veno arterial ECMO treatment and the median ECMO run was 122.7 hours. Eight survived (53.3%). LUS and CXR reached the same diagnosis in 26.6% cases. Patient with a normal LUS (A-lines) had a non lung parenchymal disease in 89% (3 Alveolar Capillary Displasia, 3 sepsis, 2 premature clousure of PDA). Comparing with the definitive diagnosis, LUS stablished the correct one in 93% cases, while CXR did in 46.6%.
Conclusions: LUS can provide additional information that helps to reach the causal diagnosis of PPHN in an early and more effective way compared to CXR being suitable for routine application in the NICU.

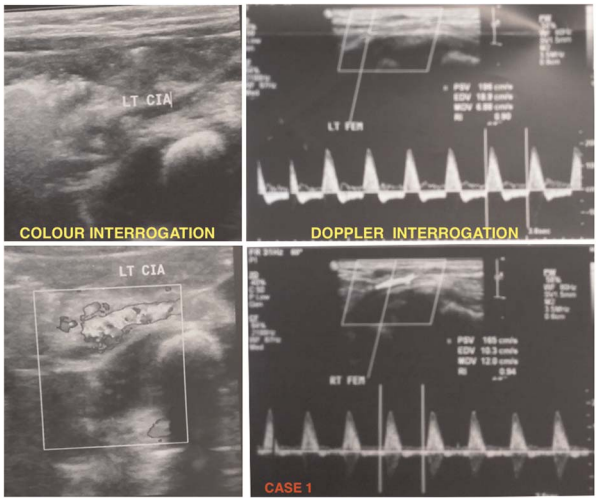

Figure 1.

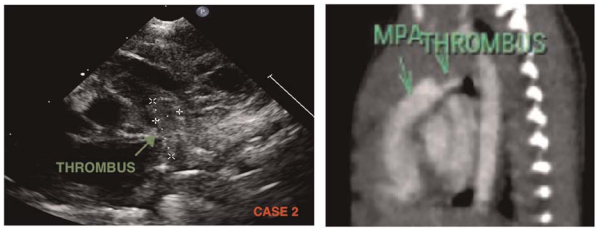

Figure 2.

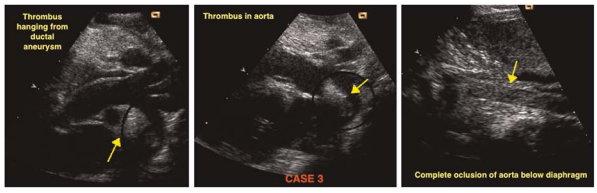

Figure 3.

P1992 - SPONTANEOUS THROMBOSIS OF GREAT VESSELS IN NEONATES WITH VARIED PRESENTATION

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Spontaneous thrombosis of great vessels is well described entity even in absence of known risk factors of sepsis, polycythemia, dehydration or thrombophilic disorders. We are sharing our experience with three neonates having unstable haaemodynamics subsequently diagnosed to have large thrombus in aorta and main pulmonary artery (MPA). Two neonates were successfully treated thrombolytic therapy while one patient succumbed due to renal failure.
Description:
Case 1: Full term neonate presented on day two of life with respiratory distress and bluish discolouration of lower limb with working diagnosis of coarctation of aorta in view of radio femoral delay and unrecordable BP in lower limb. Subsequently diagnosed to have infrarenal aortic thrombus. Successfully treated with Urokinase infusion for 72 hours followed by 4 weeks of subcutaneous low molecular weight heparin (LMWH) injections with full re-canalisation of aorta on follow up.
Case 2: One day old term baby referred for onset of breathlessness from birth. Features of right heart failure was present with low saturation of 90%. Echocardiogram revealed large thrombus in MPA extending to right and left pulmonary artery. Successfully Treated with unfractionated heparin infusion for 96 hours followed by LMWH injection for 2 weeks with complete resolution of thrombus.
Case 3: 3 days old born to diabetic mother presented in shock with history of poor oral acceptance, breathing difficulty and anuria from last 24 hours. Echocardiographic evaluation revealed biventricular dysfunction and large ductal aneurysm with thrombus insitu extending into aorta and blocking bilateral renal arteries. Baby died within one hour of admission.
Conclusion: Spontaneous thrombus involving great vessels can present with varied clinical pictures ranging from renal failure, shock, right heart failure or coarctation. High index of suspicion and basic interrogation with echocardiogram is crucial in diagnosing such conditions. Thrombolytic therapy is possible with good results even in neonates with fragile haemodynamics.

P2013 - LONGITUDINAL CHANGES IN NEAR INFRARED SPECTROSCOPY DURING THE PERI OPERATIVE PERIOD IN CHILDREN UNDERGOING FULL CORRECTION OF CYANOTIC HEART DISEASE

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Background: Changes in tissue oxygenation (rSO2) as measured by near infrared spectroscopy (NIRS) longitudinally after corrective surgery in children with cyanotic congenital heart disease (CHD) have not been previously described. We hypothesized that improvements in rSO2 occur over time as opposed to immediate

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improvement in arterial oxygen saturations as measured by pulse oximetry (SpO2). We aimed to describe the rSO2 trend before, during and after surgery in children with cyanotic CHD.

Methods: This is a prospective cohort study of children with cyanotic CHD who underwent corrective surgery. Continuous cerebral and somatic NIRS measurements were taken pre-, intra, and post-operatively for 48 hours and before hospital discharge. The Wilcoxon rank-sum test was used to compare medians of rSO2 before and after surgery, and at hospital discharge. NIRS sensors were supplied by Medtronic.

Results: Fifteen consecutive patients were recruited (Table 1). The most common lesion in our cohort was Tetralogy of Fallot in 7/15 (47%) patients. Overall mortality was 1/15 (7%). Post-operative increases in SpO2 were immediate following corrective surgery and remained in the normal range through hospital discharge (Table 2). Following corrective surgery, median post-op somatic rSO2 (%) was 80.4 compared to pre-op 74.3 ($p = 0.004$); median post-op cerebral rSO2 was 68.7 compared to pre-op 63.7 ($p = 0.636$). Median discharge levels for both somatic and cerebral rSO2 fell below pre-surgery levels: median cerebral rSO2 pre-op was 63.7 vs 60.5 at discharge ($p = 0.101$), and median somatic rSO2 was 74.3 pre-op vs 66.9 at discharge ($p = 0.064$), although differences were not statistically significant for either.

Conclusion: This pilot study observed a lack of improvement from pre-surgery levels in rSO2 following corrective surgery and suggests that targeting higher than baseline rSO2 values after surgery may not be physiological. Further studies are required to validate our findings showing lack of improvement in rSO2 after corrective surgery in children with cyanotic CHD.

Table 1. Summary of patient characteristics.

Demographic and Clinical	**All patients (n = 15)**
Age, months	18.0 (10.0, 51.0)†
Weight, kg	10.5 (7.0, 14.1)
Male gender	6 (40)††
Presence of comorbidities	3 (20)
RACHS score	
2	7 (47)
3	8 (53)
PIM 3	0.1 (0.1, 0.2)
PELOD 2	7.0 (5.0, 11.0)
Surgical	
Cardiopulmonary bypass time, minutes	193 (134, 269)
Aortic cross clamp time, minutes	110 (65, 171)
Duration surgery	335 (281, 428)

† Median (interquartile range).
†† Count (%).

Table 2. Near infrared spectroscopy O2 values† in the peri-operative period and at discharge.

	Time Periods			**Signed Rank p-values**		
NIRS Variable	**Pre-op**	**Post-op**	**Discharge**	**Pre- vs Post-op**	**Pre-op vs Discharge**	**Post-op vs Discharge**
SpO2	82.5 (76.0, 86.0)	99.8 (98.0, 100.0)	96.7 (92.3, 98.2)	0.0005	<.0001	0.0005
Cerebral rSO2	63.7 (57.2, 76.3)	68.7 (56.3, 74.8)	60.5 (53.6, 62.9)	0.6355	0.1099	0.0302
Somatic rSO2	74.3 (67.0, 81.2)	80.4 (72.0, 92.1)	66.9 (57.3, 76.9)	0.0043	0.0637	0.0043

†Median (interquartile range).

P2044 - SEVERE SEPTIC SHOCK SUCCESSFULLY MANAGED WITH ECMO IN A CHILD WITH HEMATOLOGIC MALIGNANCY

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This is a case of a 2 year-old male who was diagnosed of AML, M5 at his age of 11 months. He presented with leukocytosis and bicytopenia with atypical cells of inderminate nature about 72%. Fluorescence in situ hybridization revealed negative for MLL gene rearrangement with t(9:11)(p22:q23) on karyotypic analysis at the time of diagnosis. Under diagnosis of acute myelocytic leukemia (AML) M5, chemotherapy was started with AML 2012 protocol and achieved the complete remission (CR). However, CNS relapse occurred and at age of 16 months, unrelated peripheral blood stem cell transplantation (uPBSCT) was performed. At age of 21 months and 2 months ago, he had his 3rd relapse of AML and he underwent 2nd AML 2012 protocol of chemotherapy which also achieved complete remission. However, he had diarrhea continuously after the chemotherapy, perineal skin infection was appeared which was aggravated to abscess with necrosis of the skin and he underwent surgical debriment of the abscess. Two days after the debridment of the necrotic tissue, acute respiratory failure with hypotension developed. He underwent venoarterial extracorporeal membrane oxygenation to manage the septic shock with acute respiratory distress syndrome. After 8 days of ECMO support, he could wean off the ECMO and extubated next day. He completely recovered from septic shock and ARDS and he was discharged home after 3 months when his skin defect was completely recovered. For follow up of 5 months, he is under good condition with sustaining CR. Our case, a child with relapsed AML, previous uPBSCT with acute septic shock and acute respiratory failure, can also have many controversies, but physicians and statistics cannot always predict the outcomes of severely ill children with malignancy. ECMO should be considered in case-by-case.

P2046 - ECHOCARDIOGRAPHY IN PERSISTENT PULMONARY HYPERTENSION OF THE NEWBORN MANAGEMENT

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Background: Persistent Pulmonary Hypertension of the Newborn (PPHN) is a common neonatal problem with significant morbidity and mortality. Echocardiography is used to rule out cyanotic congenital heart disease (CHD) and to measure pulmonary vascular resistance, cardiac function and intra-cardiac shunts. The aim of this audit was to assess the management and outcomes of PPHN

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in a tertiary neonatal intensive care unit (NICU) to then develop a standard echocardiography protocol.
Materials and Methods: A retrospective chart review was conducted January 2010 - December 2015. All patients admitted to the NICU at a tertiary paediatric hospital in Perth, Australia, and requiring inhaled nitric oxide (iNO) were eligible for inclusion. Exclusion criteria included birth gestation <32 weeks and age >5 days on iNO commencement. Clinical information including cause for PPHN; ventilation type and duration; oxygenation index prior to iNO; duration of iNO; use of inotropes, Prostaglandin-E1, or Extracorporeal Membrane Oxygenation (ECMO); and mortality were recorded. The timing of the initial echocardiogram was reviewed, along with the number of subsequent studies.
Results: A total of 104 neonates were eligible for inclusion. Patient information is listed in Table 1. Full echocardiographic assessment was performed in 95/104 (91.3%), with 40/95 (42.1%) prior to iNO commencement. It was hypothesised that patients with expedient iNO commencement had severe PPHN, yet no significant difference between groups was identified (Table 2). Follow-up echocardiography was performed in 37/104 (35.6%) patients.
Conclusions: Over 90% of our cohort were assessed with echocardiography during PPHN management. Less than half of the cohort had an echo prior to iNO commencement, and only one third had a follow-up echo. Collaboration with the NICU is required to ensure a comprehensive echocardiogram is performed in hypoxic patients prior to iNO to rule out CHD. Subsequent echo assessment may be targeted to assess response to therapy.

Table 1. Clinical features and outcomes.

Median Gestational Age	38 + 6
Median Birth Weight	3260 g
Diagnoses	Meconium aspiration n = 36 Hyaline Membrane Disease n = 24 Hypoxic Ischaemic Encephalopathy n = 16
Mean ventilation hours	147 (range 7-622)
High frequency oscillation ventilation	46 (44.2%)
Oxygenation index >20	36 (34.6%)
Mean iNO hours	82 (range 1-289)
Inotropic agents	72 (69.2%)
Prostaglandin-E1	24 (23.1%)
ECMO	7 (6.7%)
Death	13 (12.5%)

Table 2. Clinical features and outcomes based on timing of echocardiogram.

	Echocardiogram pre-iNO (n = 40)	Echocardiogram post-iNO (n = 55)	p value
Mean ventilation hours	179 (range 29-579)	128 (range 7-622)	0.647
High frequency oscillation ventilation	19 (47.5%)	26 (47.3%)	1.000
Oxygenation index >20	15 (37.5%)	21 (38.2%)	1.000
Mean iNO hours	83 (range 1-280)	85 (range 7-289)	0.9096
Inotropic agents	27 (67.5%)	45 (81.8%)	0.1459
Prostaglandin-E1	9 (22.5%)	15 (27.3%)	0.6401
ECMO	4 (10%)	3 (5.5%)	0.4495
Death	3 (7.5%)	10 (18.2%)	0.2258

P2061 - TITLE SUDDEN CARDIAC DEATH IN CHILDREN UNDER 14 YEARS OF AGE IN OUR ZONE

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Matherial and Methods: We performed a retrospective study, collecting clinical and autopsy information on SCD cases with cardiac cause among children 0 to 14 years of age in Gipuzkoa (Spain) from 2005 through 2016. When no previous cardiac cause justifying cardiac death was known, autopsy and genetic testing was offered to relatives.
Results: 16 cases of SCD with cardiac cause were identified. The anual incidence was 1,1 cases per 100000 persons 1 to 14 years of age. 63% of the cases involved men. Median age of cardiac death event was 5,9 months (inter-quart rank of 58,3 months). The most common explained causes of cardiac sudden death were primary pulmonary hypertension in syndromic patients (3 cases; 18,7%), coronary artery anomalies (2 cases; 12,5%), long QT syndrome (2 cases; 12,5%), acute myocarditis (2 cases; 12,5%), bacterial endocarditis (1 case; 6,3%), ventricular fibrillation (1 case; 6,3%) and irreparable structural congenital heart disease (5 cases 31,3%; 2 term newbors: 1 with hypoplastic pulmonary veins and 1 massive systemic-to-pulmonary venous fistulae; and 3 extremely preterm infants: 1 abnormal pulmonary venous return, 1 aortic coarctation and 1 critical pulmonary stenosis). 10 cases (62,5%) had previous diagnosis of primary cardiac anomalies, 3 cases (18,7%) had autopsy findings of acquired cardiac anomalies and 3 cases (18,7%) were diagnosed after successful cardiopulmonary resuscitation (CPR) by cardiological studies and/or genetic testing. 3 cases (18,7%) recovered from SCD after resuscitation with no significant sequel. 7 cases (43,8%) were outpatient SCD, 5 of them (71,4%) were witnessed and received advanced CPR, but only 2 (28,6%) of them reverted (60% death risk even with early CPR onset (95% Confidence Interval: 0,29-1,23).
Conclusion: Autopsy and genetic investigation increases SCD cause identification. Witnessed SCD and early CPR is the key to its recovery.

P2082 - FETAL VERSUS POSTNATAL DIAGNOSIS OF INTERRUPTED AORTIC ARCH AND ITS IMPACT IN SHORT AND LONG TERM OUTCOMES

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Background: Prenatal diagnosis of interrupted aortic arch (IAA) allows for comprehensive fetal and genetic counseling, but the impact on post-natal outcomes remains unknown. Our study aims to evaluate the impact of fetal (FD) versus post-natal (PND) diagnosis of IAA on pre and post-surgical outcomes.
Methods: A retrospective review of confirmed IAA patients, from 2004 to 2016 was performed. Patients with a lethal genetic disorder (i.e.: trisomy 13,18), severe extra-cardiac malformations, pre-term <34 weeks, birth weight less than 2.5 kg and patients to receive compassionate care were excluded. Patients were grouped as FD or PND based on timing of the initial suspicion of a

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prostaglandin (PGE) dependent lesion. Patients were also grouped according to the presence of usual (i.e.: VSD) versus complex associated lesions (CAL; i.e.: truncus arteriosus) and if genetic disorders (GD) were present. Pre and post-surgery clinical data and long-term arch geometry were documented. Survival and outcome analysis were performed.

Results: Twenty-one fetal and 39 postnatal infants with IAA were analyzed. There were similar distributions of gender, gestational age at birth and IAA type in both groups (Table). There was a decreased frequency of 22q11 microdeletion in FD 9.5% vs PND 48.7%, p = 0.004. Fetal group was associated with more complex lesions (CAL: FD 57.1% vs PND 17.9%, p = 0.003). The FD group needed lower doses of PGE, shorter time of mechanical ventilation and ICU/hospital stay prior to surgical repair. Surgery occurred earlier in the FD group (FD at 3 (range 2-6) days, PND 7 (3-77) days, p = 0.001). There were no differences in post-operative mortality, hemodynamic or respiratory support or long-term arch geometry. Similar findings were observed after adjusting for the presence of GD or CAL.

Conclusions: Fetal diagnosis of IAA improves the pre-surgical outcomes; however has no impact on post-operative outcomes or on long-term aortic arch geometry.

Table 1. Demographic, clinical, pre-surgical, surgical, post-operative and long term data according to fetal or post-natal diagnosis of PGE dependent lesion. Data presented as: Mean ± standard deviation, median (range) or n (%) as appropriate.

	All patients			Excluding genetic syndromes		
	Fetal	**Post-natal**	**p**	**Fetal**	**Post-natal**	**p**
Number of cases	21	39		14	16	
Gender: Male (%)	14 (66.7%)	24 (61.5%)	0.78	10 (71.4%)	9 (56.3%)	0.47
Weight at birth, kg	3.5 ± 0.5	3.2 ± 0.4	0.06	3.5 ± 0.7	3.2 ± 0.4	0.24
Gestational age at birth, weeks	38.6 ± 1.1	38.4 ± 1.4	0.61	39 ± 1	38.7 ± 1.3	0.57
Presence of a genetic disorder	**5 (23.8%)**	**23 (59%)**	**0.008**	-	-	
22q11.2 (%)	**2 (9.5%)**	**19 (48.7%)**	**0.004**	-	-	
Presence of CAL (%)	12 (57.1%)	7 (17.9%)	0.003	8 (57.1%)	5 (31.3%)	0.27
Truncus arteriosus (%)	3 (14.3%)	5 (12.8%)	1	3 (21.4%)	4 (25%)	1
TGA physiology (%)	**4 (19%)**	**1 (2.6%)**	0.047	2 (14.3%)	1 (6.3%)	0.59
Single ventricle (%)	**4 (19%)**	**0**	0.01	3 (21.4%)	0	0.09
Other CHD (%)	1 (4.8%)	2 (5.1%)	1	0	1 (6.3%)	1
Type of IAA			0.25			
A (%)	10 (47.6%)	11 (28.2)	0.16	7 (50%)	9 (56.3%)	1
B (%)	11 (52.4%)	27 (69.2)	0.26	7 (50%)	7 (43.8%)	1
C (%)	0	1 (2.6%)	1	0	0	
PRE- SURGICAL AND SURGICAL DATA						
Death before surgery	0	1 (2.6%)	1	0	0	1
PGE start day (of life)	**0 (0 - 1)**	**2 (0 - 31)**	**0.001**	**0 (0 - 1)**	**3 (0 - 31)**	**0.001**
Days of PGE	**3 (2 - 6)**	**5 (1 - 72)**	**0.001**	3 (2 - 6)	5 (1 - 72)	0.22
Maximal dose of PGE: mcg/kg/min	0.01 (0.01 - 0.05)	0.05 (0.01 - 0.2)	**0.001**	0.01 (0.01 - 0.05)	0.02 (0.01 - 0.2)	**0.043**
Mech. ventilation pre-surgery (%)	**5 (23.8%)**	**21 (53.8%)**	**0.03**	3 (21.4%)	6 (37.5%)	0.44
Days of mech. ventilation pre-surgery	**0 (0 - 4)**	**1 (0 - 23)**	**0.007**	0 (0 - 4)	0 (0 - 12)	0.38
Maximal lactate, mmol/L	**4.6 ± 2.3**	**9.8 ± 6.9**	**0.01**	-	11.2 ± 7.6	0.53
Inotropic support pre-surgery (%)	5 (23.8%)	12 (30.8%)	0.77	3 (21.4%)	3 (18.8%)	1
Days of ICU/CCCU stay pre-surgery	**3 (1 - 5)**	**5 (0 - 18)**	**0.001**	**3 (1 - 5)**	**5 (0 - 18)**	**0.02**
Days of hospital stay pre-surgery	**3 (2 - 6)**	**6 (2 - 18)**	**0.001**	**3 (2 - 6)**	**5.5 (3 - 18)**	**0.001**
Age at surgery (days)	**3 (2 - 6)**	**7 (3 - 77)**	**0.001**	**3 (2 - 6)**	**9 (3 - 77)**	**0.001**
POST- OPERATIVE DATA						
Death after surgery (%)	1 (4.8%)	2 (5.1%)	1	1 (7.1%)	1 (6.3%)	1
Days of inotropic support	6 (0 - 208)	7 (1 -91)	0.86	6.5 (0 - 208)	6 (1 - 68)	0.89
Days of mech. ventilation	7 (1 - 135)	5.5 (1 - 91)	0.12	6.5 (1 - 135)	5 (1 - 91)	0.29
Days of ICU/CCCU stay post-surgery	9 (2 - 52)	8 (2 - 91)	0.31	8 (2 - 52)	8.5 (2 - 91)	0.82
Total length of stay	28 (7 - 212)	32 (8 - 105)	0.52	21 (7 - 212)	34.5 (10-105)	0.31
LONG TERM FOLLOW-UP						
Death after discharge	1 (4.8%)	0	0.35	0	0	
Last available echocardiogram						
Age at echocardiogram, years	2.8 ± 3.4	3.94 ± 3.93	0.32	3.1 ± 3.9	3.6 ± 3.7	0.77
BSA, m^2	0.64 ± 0.25	0.66 ± 0.26	0.84	0.74 ± 0.23	0.69 ± 0.26	0.75
Aortic valve Z-score	2.34 ± 4.57	0.13 ± 3.57	0.16	3.89 ± 6.0	1.44 ± 4.22	0.39
Ascending Aorta Z-score	1.73 ± 1.63	1.80 ± 2.2	0.95	1.95 ± 1.53	3.07 ± 2.37	0.55
Transverse arch Z-score	3.37 ± 1.93	2.72 ± 2.22	0.46	3.32 ± 0.59	2.54 ± 2.69	0.54
Isthmus Z-score	1.11 ± 1.59	0.82 ± 1.66	0.68	0.90 ± 1.80	0.07 ± 1.29	0.32
Descending aorta Z-score	1.67 ± 1.66	0.99 ± 1.48	0.29	1.61 ± 1.88	0.34 ± 1.11	0.12

	Excluding CAL		
	Fetal	**Post-natal**	p
Number of cases	9	32	
Gender: Male (%)	7 (77.8%)	21 (65.6%)	0.69
Weight at birth, kg	3.1 ± 0.3	3.2 ± 0.4	0.53
Gestational age at birth, weeks	38.4 ± 0.9	38.5 ± 1.4	0.97
Presence of a genetic disorder	3 (33.3%)	21 (65.6%)	0.13
22q11.2 (%)	2 (22.2%)	17 (53.1%)	0.2
Presence of CAL (%)	-	-	
Truncus arteriosus (%)	-	-	
TGA physiology (%)	-	-	
Single ventricle (%)	-	-	
Other CHD (%)	-	-	
Type of IAA			0.76
A (%)	3 (33.3%)	8 (25%)	0.68
B (%)	6 (66.7%)	23 (71.9%)	1
C (%)	0	1 (3.1%)	1
PRE- SURGICAL AND SURGICAL DATA			
Death before surgery	0	1 (3.1%)	1
PGE start day (of life)	**0 (0 - 0)**	**2 (0 - 10)**	**0.001**
Days of PGE	**3 (2 - 5)**	**5 (2 - 72)**	**0.001**
Maximal dose of PGE: mcg/kg/min	0.01 (0.01 - 0.05)	0.05 (0.01 - 0.2)	**0.005**
Mech. ventilation pre-surgery (%)	2 (22.2%)	20 (62.5%)	0.057
Days of mech. ventilation pre-surgery	**0 (0 - 1)**	**2 (0 - 23)**	**0.02**
Maximal lactate, mmol/L	**3.6 ± 1.7**	**11 ± 6.9**	**0.003**
Inotropic support pre-surgery (%)	2 (22.2%)	12 (37.5%)	0.69
Days of ICU/CCCU stay pre-surgery	**3 (2 - 4)**	**5 (0 - 18)**	**0.001**
Days of hospital stay pre-surgery	**3 (2 - 4)**	**6 (2 - 18)**	**0.001**
Age at surgery (days)	**3 (2 - 5)**	**8 (4 - 77)**	**0.001**
POST- OPERATIVE DATA			
Death after surgery (%)	0	1 (3.1%)	1
Days of inotropic support	5 (1 - 12)	6 (1 - 68)	0.35
Days of mech. ventilation	6 (1 - 42)	5 (1 - 91)	0.85
Days of ICU/CCCU stay post-surgery	8 (2 - 52)	8 (2 - 91)	0.68
Total length of stay	21 (11 - 120)	33 (8 - 105)	0.14
LONG TERM FOLLOW-UP			
Death after discharge	0	0	
Last available echocardiogram			
Age at echocardiogram, years	3.0 ± 3.9	3.5 ± 3.8	0.75
BSA, m^2	0.58 ± 0.25	0.62 ± 0.24	0.79
Aortic valve Z-score	-0.24 ± 4.72	-0.84 ± 0.24	0.7
Ascending Aorta Z-score	1.64 ± 1.98	1.14 ± 1.59	0.64
Transverse arch Z-score	4.34 ± 1.33	2.84 ± 2.15	0.15
Isthmus Z-score	1.94 ± 1.31	1.08 ± 1.62	0.28
Descending aorta Z-score	2.45 ± 1.66	1.11 ± 1.53	0.1

Abbreviations (not mentioned in the abstract): BSA: Body surface area; ICU: Intensive care unit; CCCU: Cardiac critical care unit; TGA: Transposition of the great arteries;

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P2086 - EFFECTIVENESS OF A PROGRAM TO REDUCE SURGICAL SITE INFECTION IN CONGENITAL HEART SURGERY IN A REFERAL PEDIATRIC HOSPITAL IN ARGENTINA

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Introduction: Surgical site infections (SSI) in congenital heart surgery are an important cause of morbidity and mortality, and use of hospital resources. In the context of an increasing SSI rate we developed an intervention program to reduce SSI.
Objective: Describe the results of a multidimensional intervention program to reduce SSI in a congenital heart surgery program.
Setting: operating room and pediatric cardiac intensive care unit at Hospital de Pediatria "J.P. Garrahan" Intervention: Professional supervision of preoperative bath with 4% chlorhexidine soap, incorporation of 2% alcoholic chlorhexidine for skin antisepsis, educational workshops, and an infection control nurse present in the operating room area.
Patients and Methods: all patients undergoing cardiac surgery performed between 1/1/2015 to 12/31/2016 were included. SSI rates pre and post intervention were compared. A subgroup analysis was performed in patients with delayed sternal closure (STATA 9.0).
Results: 1145 surgeries were performed, 14% with delayed sternal closure. Adequate preoperative antibiotic prophylaxis was confirmed in at least 881 patients (77%). 58 infections were observed (5.06%), 60% superficial, and 40% deep/mediastinitis. Bacterial isolation was 64% gram + cocci, 21% gram -, and 16% enterobacteriaceae. After intervention we observed a reduction in the infection rate from 4.1% to 2.41% with RR 1.25 (95% CI 0.97-1.62, p 0.13) in patients with primary sternal closure. This reduction was observed for both superficial 16 (76.2%) vs 9 (81.8%) and deep infections 5 (23.8%) vs 2 (18.1%). In patients with delayed chest closure no SSI reduction was found 13.7% vs. 14.1%.
Conclusion: The program was useful, a reduction in the SSI rate was observed in patients with primary chest closure. Infection rate remained high in children with delayed sternal closure. The program and epidemiological surveillance should be continued to improve the quality of care for children undergoing cardiovascular surgery.

P2089 - WEBINARS' BASED COURSE IN PEDIATRIC CARDIAC INTENSIVE CARE UNIT A STRATEGY TO IMPROVE NURSES' CONTINUOUS EDUCATION

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Introduction: Quality Improvement Collaborative for Congenital Heart Surgery (IQIC) webinars are a valuable educational resource for staff involved in the care of patients with congenital heart disease. However in our environment there are barriers to optimum utilization. To promote continuous education we designed a virtual course for pediatric cardiac intensive care unit (PCICU) nurses.
Objective: to describe a continuous training course for the PCICU nursing staff.
Setting: Garrahan Pediatric Hospital. Buenos Aires. Argentina
Material and Methods: With IQIC authorization a continuous virtual training course was designed for the PCICU staff. This course was mandatory for new nurses, optional for rest of the staff and grants credits for institutional professional promotion. 10 translated webinars were selected considering staff training priorities, complemented with additional audiovisual material and suggested readings, 4 workshops and 3 in situ simulation scenarios. Partial assessments and final examination was required for approval. A satisfaction survey was completed at the end of the course.
Results: Fifty nurses (56% of the total staff) were enrolled: 80% with master degreeand 20% registered nurses. Most of them (60%) with more than 10 years working in the CICU, while 20% were recently hired. 48 nurses (96%) completed the course and approved exams. Satisfaction survey analysis showed 10% very satisfied, 80%satisfied, and only 10% unsatisfied.
Conclusions: The course was very successful, more than half of PCICU nursing staff enrolled, complete and approved the course. Ninety percent were satisfied or very satisfied with it. This new training tools are very useful in professional continuous education in our unit.

P2105 - NEONATAL DEATH ARISING FROM CONGENITAL HEART DISEASE IN A TERTIARY NICU

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Introduction: Congenital heart disease (CHD) is responsible for more deaths in the first year than any other birth defects. The causes and circumstances of neonatal death have barely been studied in these patients.
Aim: To examine causes and manner of death, and characteristics of the dying process in patients with CHD, in our NICU.
Method: observational retrospective study. Study group: deceased patients with CHD from January 2009-December 2016. Medical histories were checked and epidemiological and characteristics of the dying process were registered.
Results: During the period of study there were 5331 admissions (716 patients with CHD) and 283 deceased patients. 61/283 (21.5%) of deceased patients had a CHD. Average gestational age in CHD was 37.4w, weight 2700 g, 52.4% were males. 31.1% were outside admissions. Age at the moment of death: 18.5 days. Causes of death : 1) somatic poor prognosis (not candidate for surgery)(21/61), 2) a context of multiple malformation syndrome and/or chromosome abnormalities (20/61) and 3) complications related to surgery (18/61). Some types of CHD: 20 SCIH or obstruction of the left heart outflow tract, 6 complex TGV, 2 total APVR, 6 complex CHD (right and left side affected). Treatment limitation (TL) was settled in 37/61(60.7)% and in 83% of the patients with CHD associated to multiple malformation syndromes and/or chromosome abnormalities . Types of TL: withdrawing life-support (73% of cases and withholding new treatments: 27%. All the patients that died after TL were accompanied by their parents. Autopsy was granted in 52.5%.
Conclusions: The most significant groups of newborns with CHD who die during admission to NICU are those that have a poor somatic prognosis and those that link multiple malformation syndromes and/or chromosome abnormalities. The majority die after TL, predominantly after life support is withdrawn.

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P2106 - PROMOTING COLLABORATION BETWEEN NURSES AND PHYSICIANS IN PAEDIATRIC CARDIAC INTENSIVE CARE CHARACTERISING INTERPROFESSIONAL RELATIONSHIPS ATTITUDES AND BEHAVIOURS

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Background: The paediatric cardiac intensive care unit (CICU) is a 'high-stakes' environment in which inter-professional relationships may come under great pressure. Collaboration between nurses and physicians improves patient safety and job satisfaction whilst reducing burnout and staff turnover (1,2). The aims of this study were: to characterise the perceptions of nurse-physician relationships in our paediatric CICU (a 21-bed tertiary/quaternary centre); to identify strategies to promote collaboration; and to obtain 'benchmark' data for assessing the efficacy of these strategies going forward.
Methods: Anonymous electronic survey of medical and nursing staff working on CICU during May-November 2016.
Results: •90/168 (54%) surveys were completed; 71/124 (57%) from nurses and 19/44 (43%) from physicians. •90% rated the overall quality of nurse-physician relationships on CICU as 'somewhat', 'very' or 'extremely' positive. •85% reported positive interactions occurring daily; examples included shared decision making/learning and mutual respect. •Negative interactions occurred less frequently (5% daily; 33% 2-weekly; 19% monthly); examples included: not introducing oneself; dismissing clinical concerns; and comments perceived as patronising and/or disrespectful. • > 50% rated the following as 'absolutely critical' in influencing nurse-physician relationships: respect; communication/listening skills; and common goals. •Less influential, but still important factors included: consultant/nurse in charge and patient workload. • > 90% reported joint ownership of the patient with 70% agreeing that nurses and physicians shared decision making. •Respondents indicated that beneficial strategies for improving relationships might include: joint simulation/ teaching sessions (>90%); social functions (88%); and a nurse-physician 'buddy' system (72%).
Conclusions: This study suggested a predominance of positive inter-professional interactions/relationships on our unit; however, areas for improvement were identified. Several potential interventions for improving relationships emerged, including simply presenting these data to new starters to encourage reflection and improve awareness. The data could also be used as a benchmark for future interventions.
References: 1) Baggs JG et al (1999). Association between nurse-physician collaboration and patient outcomes in three intensive care units. Crit Care Med 27:1991-1998. 2) Vahey DC et al (2004). Nurse burnout and patient satisfaction. Med Care 42:1157-1166.

P2118 - PULMONARY LYMPHANGIECTASIA A RARE BUT SERIOUS OUTCOME PREDICTOR IN NEONATES WITH TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE

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Background: Congenital Pulmonary Lymphangiectasia (CPL) is a very rare entity. Type 2 CPL has been noted to be associated with congenital heart diseases such as TAPVD. Most patients with CPL tend to have a poor prognosis with significant mortality being reported. There is very little information on the clinical characteristics including morbidity and mortality in this subset.
Methods: Review of all patients diagnosed with TAPVD between 2012 and 2016 in a tertiary care hospital in Singapore. Patients with complex heterotaxy syndromes were excluded. Patients with biopsy proven pulmonary lymphangiectasia were studied.
Results: 15 patients with TAPVD were identified. In all patients, the diagnosis was confirmed postnatally by echocardiogram. Five patients (33%) had obstruction to the pulmonary venous drainage in our series; 2 of 5 (40%) were identified with pulmonary lymphangiectasia through CT imaging and confirmed by histopathology. There were 7 patients (47%) with supracardiac TAPVD and 2 had obstructed drainage; 4 patients (27%) had a cardiac type TAPVD and none were obstructed; 2 patients (13%) had mixed type TAPVD and 2 patients (13%) had Infracardiac TAPVD. Both patients with Infracardiac TAPVDs and 1 patient in mixed type with partial infradiaphragmatic drainage were considered to be obstructed physiologically, even if Doppler studies were negative. One patient with lymphangiectasia with supracardiac obstructed TAPVD succumbed by postoperative day 82 despite correction of TAPVD on Day 1 of life. The other patient with pulmonary lymphangiectasia and Infracardiac TAPVD survived till date despite later repair (day 18 of life). Both required prolonged and difficult ventilation post surgery.
Conclusions: 1. Patients with TAPVD and pulmonary lymphangiectasia are complicated by prolonged ventilation even if corrective repair is done. 2. Presence of pulmonary lymphangiectasia may predict higher mortality but certainly portends significant morbidity compared to isolated TAPVD. 3. Pulmonary lymphangiectasia can occur irrespective of the type of TAPVD.

P2125 - PULMONARY ARTERY HYPERTENSION MANIFESTING AS SYNCOPE WITH CYANOSIS. A CASE REPORT

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A 15 month old female child, weight 5.7 kg referred with a history of sudden loss of consciousness with intermittent cyanosis after episodes of excessive irritability, for previous 3 months. The patient was the second of twin preterm delivery (28 weeks gestation), small for gestational age (SGA) with birth weight 700 gms. The patient had prolonged NICU stay and underwent PDA ligation due to inability to wean off non invasive ventilation. The patient was referred to a neurophysician and was investigated for the same, but the imaging and electroencephalogram (EEG)were normal. A 2D echocardiography with color Doppler revealed the diagnosis as severe right upper pulmonary vein discrete stenosis with a mean gradient of 8 mm hg with bidirectionally shunting secundum atrial septal defect (ASD) with severe tricuspid regurgitation (TR) and severe pulmonary hypertension (PAH), confirmed with CT pulmonary angiography. The child was taken for a high risk surgical intervention (Sutureless neoatrium technique) with repair of pulmonary vein stenosis with extrapericardial cannulation and fenestrated ASD closure. The patient required prolonged ventilation due to recurrent desaturation with hemodynamic instability with systemic PAH postoperatively and required inodilators with oral pulmonary vasodilators at maximum

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dosage including intravenous sildenafil infusion for reducing PAH. The patient could only be extubated by the 8th post operative day (POD) on nasal continuous positive airway pressure (CPAP), which was withdrawn on the 11th POD. The patient on discharge had half systemic PAH and maintained saturation greater than 90% on room air. On one month follow up the PAH had regressed to one third systemic and the patient had improved appetite with weight gain and no recurrence of syncope. To conclude syncope with cyanosis in younger children is a rare manifestation of PAH and warrants timely intervention.

P2126 - PREDICTORS OF PROLONGED MECHANICAL VENTILATION PROLONGED INTENSIVE CARE UNIT STAY AND MORTALITY IN PEDIATRIC CONGENITAL CARDIAC SURGERY PATIENTS IN INDIAN POPULATION

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Background: Knowledge of factors predicting prolonged mechanical ventilation (MV), ICU stay and mortality in children undergoing cardiac surgery can help in early treatment and targeted therapy. We aimed to determine such predictors in pediatric patients undergoing surgery for congenital heart disease in a tertiary hospital.
Methods: This ongoing prospective observational study has enrolled 34 patients till now. Perioperative data including hematological and biochemical parameters, cardiopulmonary bypass(CPB) time, CPB temperature, aortic cross clamp time, blood gas analysis, lactate level, drain output, transfusion data, Vasoactive-inotropic score (VIS), renal score, ventilation index (VI), Vasoactive-ventilation-renal (VVR) score are recorded pre-operatively, at the time of receiving in ICU and 6, 24 and 48 hours postoperatively.
Results: Median age and weight were 12 months (IQR: 3 -36) and 6.5 kg (IQR: 4-11) respectively. Median mechanical ventilation duration and ICU stay were 50 hours (IQR: 17-120) and 7 days (IQR: 4-10) respectively. Lower age, prolonged CPB time, Low platelet counts and high renal index on receiving in ICU, high renal index, high bilirubin and low pao2 at 6 hours, high creatinine, increased creatinine, high lactate, high ventilation index and high VVR score at 48 hours in ICU were all independently associated with prolonged mechanical ventilation. Similarly higher intraoperative base deficit, high VIS score at 30 minutes post CPB, lower venous CO2 on receiving in ICU, high VIS score and high VVR after 6, 24 and 48 hours in ICU, low arterial CO2 at 48 hour in ICU were independently associated with prolonged ICU stay. Low pH in the early postoperative period and increased 2 day drain output were significantly associated with high mortality.
Conclusion: While patients with abnormal renal parameters had prolonged mechanical ventilation, higher inotrope requirement patients had a longer ICU stay. Acidosis and mortality went together in this interim analysis.

P2129 - ASSOCIATION OF HIGH VASOACTIVE-INOTROPIC SCORE WITH POOR OUTCOMES AFTER PEDIATRIC CARDIAC SURGERY IN INDONESIA

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Background: The use of high vasoactives on leaving operating room was associated with poor outcomes in children after cardiac surgery. The more severe the hemodynamic state, the lower the cardiac index, and therefore, a higher vasoactive inotropic support were needed. These might complicate to death.
Objective: To determine the association of high vasoactive-inotropic score with poor outcomes in children after cardiac surgery in Indonesia.
Methods: A prospective cohort study was conducted from April 2014 to March 2015 on all children undergoing cardiac surgery at Dr Cipto Mangunkusumo Hospital, Jakarta, Indonesia. The use of vasoactive agents at cardiac ICU was recorded for the first 24 hours after cardiac surgery. The use of vasoactive support and its association with poor outcomes including in-hospital mortality, cardiac arrest, and multi-organ failure were analyzed. Multivariable logistic regression analyses were used to quantify the association between high vasoactive-inotropic score and poor outcomes allowing for statistical control of age, sex, nutritional state, syndrome, cardio-pulmonary bypass, and complexity of surgery. High vasoactive-inotropic score was defined as more than or equal to 20. Formula of vasoactive-inotropic score was dopamine dose + dobutamine dose + (100 x epinephrine dose) + (10 × milrinone dose) + (100 × norepinephrine dose). Doses were in μg/kg/minutes.
Results: We recruited 257 patients with median age of 36 months. Of those, 32 (12.5%) were administered to a high vasoactive-inotropic score. Median vasoactive-inotropic score was 10 (3–42). After adjusting for patient characteristics and the complexity of surgery; there was a significant association between high vasoactive-inotropic score and poor outcomes in children after cardiac surgery including mortality (odds ratio 56.3; 95% CI 17.9–176.8), cardiac arrest (odds ratio 5.6; 95% CI 1.6–19.6), and multi-organ failure (odds ratio 17.8; 95% CI 5.6–56.3).
Conclusions: High vasoactive-inotropic score is significantly associated with in-hospital mortality and other poor outcomes in children after cardiac surgery.

P2163 - ARRHYTHMIA IN THE NEONATAL INTENSIVE CARE UNIT AT OSAKA MEDICAL COLLEGE HOSPITAL

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Objective: The aim of the present study is to analyze non benign neonatal arrhythmias (NBNA) observed in a neonatal intensive care unit (NICU) at Osaka Medical College Hospital.
Materials and Methods: From January 2010 to December 2016, newborns admitted to our NICU due to NBNA were evaluated retrospectively. We defined the arrhythmias such as supraventricular tachycardia (SVT), sinoatrial node dysfunction, disorders of the atrioventricular (AV) conduction system, atrial flatter (AF), ventricular tachycardia (VT), long QT syndrome, and ventricular fibrillation (VF) which need treatment or long term follow up as NBNA.
Results: Fourteen cases were included in this study. The incidence of NBNA was 1.2% of total admission of NICU. The median age at diagnosis was 10 day ranging from 0 to 187 day. Twelve of 14 patients had congenital heart disease (CHD). The most common type of congenital heart disease was total anomalous of pulmonary venous connection (TAPVC) and heterotaxy syndrome,

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following tetralogy of Fallot. In eight cases with CHD, NBNA occurred postoperative period from 1 to 61 days (median 10 days) after surgical operation. The most frequent type of arrhythmia was SVT (57%). Other types of NBNA were advanced AV block (22%), sick sinus syndrome (14%) and VT (7%). All cases but three case with AV block underwent medical treatment. Propranolol was used to treat of 6 patients with SVT (42.9%). Overall mortality rate was 35.7% (n = 5). Three of 5 deaths were associated with NBNA. All three cases associated with NBNA had advanced AV block.
Conclusion: In this study, NBNA in our NICU were associated with cardiac surgery. The most common type of arrhythmia in this study was SVT after total repair for TAPVC. Advanced AV block had poor prognosis and occurred late after cardiac surgery.

P2165 - IMPACT OF MULTIPLE LOW FLOW INTRAVENOUS INFUSIONS ON VOLUME AND DRUG DELIVERY

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Background: In practice of neonatal postoerative cardiac surgery, using low volume with multiple concurrent drug infusion via peripheral vein sometime is inevitable. These could be altered the accuracy of drug delivery. Data of accuracy for concurrent low rate fluid infusion should be demonstrated.
Objectives: To verified accuracy of infusion pumps under low infusion rate and to study accuracy of concurrent infusion at low rate.
Methods: Ten Infusion pumps and ten syringe pumps were tested for accuracy and precision of fluid delivery at multiple flow rate. The most accurate pumps type were used to deliver concurrent Copper sulfate solution rate 1 mL/h side tube, 1 mL/h of sterile water side tube and 3 mL/h main tube through a multiple stopcock connection. Samples were collected in two-minute interval over a 60 minute period to measure optical density of Copper sulfate. Then Copper sulfate concentration was calculated by using optical density-concentration curve.
Results: Mean actual rate of infusion pump at preset rate 0.1, 0.5, 1, 2, 5 mL/h are 0.1177, 0.4933, 0.9996, 1.9990, 4.9831 mL/h respectively. And mean actual rate of syringe pump at the same preset rate are 0.0114, 0.4986, 0.9811, 2.0063, 4.9948 mL/h respectively. Delivering of Copper Sulfate at main tube rate of 3 mL/h and side tube rate of 1 mL/h through the near patient port of multiple stopcock connection and far from patient port take 30 and 40 minutes respectively to reach the steady stage, and for spider triple lumen, 25 minutes is required to reach the steady stage.
Conclusions: At low flow rate of less than 2 mL/h. the infusion pump is more accurate than syringe pump. Using concurrent infusion has significant effect of early delivery precision and this should be avoid by using multiple lumen catheter for delivery of inotropic agents.

P2172 - VALUE OF NEAR INFRARED SPECTROSCOPY AS A PROXY FOR CARDIAC OUTPUT IN AN INFANT ON EXTRACORPOREAL MEMBRANE OXYGENATION A CASE REPORT

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Background: Low cardiac output syndrome after cardiopulmonary bypass (CPB) in neonates occurs in 25% and extracorporeal membrane oxygenation (ECMO) is the treatment of choice in extreme cases. Actual cardiac output is difficult to measure in children, therefore proxies like peripheral perfusion, pulses, urine output, arterial blood pressure, metabolic acidosis, lactate levels must be followed. We observed the correlation between NIRS and other proxies in relationship to cardiac output.
Material and Methods: A neonate diagnosed with single ventricle, transposition of great arteries and restrictive atrial septal defect underwent a palliative procedure: atrial-septostomy and banding of the pulmonary artery. Cerebral oxygen saturation was monitored via NIRS (INVOS ™5100C) during surgery. Severe ventricular failure with low cardiac output when attempting to wean from CPB despite maximal inotropic therapy was managed with ECMO through central cannulation. Blood pressure, NIRS levels, base deficit and urine output were followed during 96 hrs of ECMO support as proxies for cardiac output (see figure 1). After ECMO was weaned off with an inotropic support and stable blood pressures the NIRS value dropped from 55 to 20 with a lactate rise (3,69). After 40' low blood pressures followed, with severe metabolic acidosis and ST changes. Unable to go back on ECMO due to technical issues, pediatric advanced life support was unsuccessful to reverse the situation.
Results: See Figure 1
Conclusion: Near infrared spectroscopy (NIRS) had the ability to monitor and predict low cardiac output in this patient. When confronted with patients lacking direct measure of cardiac output due to weight restrictions, the use of NIRS seems to be at least as effective as other proxies utilized to that end.

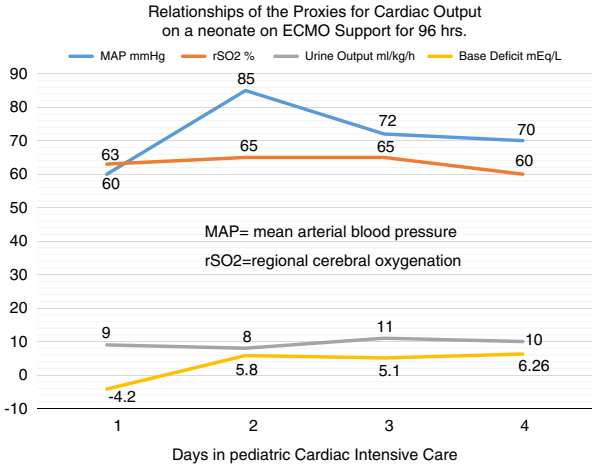

Figure.

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P2192 - RESTRICTIVE RIGHT VENTRICLE PHYSIOLOGY IN TETRALOGY OF FALLOT DURING EARLY POSTOPERATIVE PERIOD

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Background: The mortality rate of tetralogy of Fallot (TOF) decreases as the surgical experience increases and the intensive care unit (ICU) follow-up improves. In this study, the effects of restrictive right ventricle in postoperative period were investigated in TOF patients undergone total correction.
Materials and Methods: Between January 2015 and November 2016, 112 cases that were undergone total correction were included in the study. Cases are identified as restrictive right ventricle (RRV +) and the ones with peritoneal dialysis were applied (Group 1), and the ones without the restrictive right ventricle (RRV-)(Group 2) according to their antegrade pulmonary flow using pulse wave in postoperative echocardiography. The demographical datas, preoperative echocardiographic and angiographic datas, type and duration of the operation, complications in the ICU were evaluated.
Results: Median age of the cases were 12 months (range 1 month-16 years), median weight was 8.7 kg (range 3.3-41). Group 1 included 17 cases and Group 2 included 95 cases.The McGoon index, z scores of the pulmonary annulus, the pulmonary artery, RPA and LPA were similar in both groups. Intraoperative datas (cardiopulmonary bypass (CPB) time, cross clamp (CC) time, right ventricular/left ventricular pressure ratio (pRV/pLV), usage of transannular patch) were also similar. In Group 1 the incidence of residual defect and pulmonary regurgitation were higher.Also in group 1, the inotrope score, duration of stay in the ICU and in the hospital were statistically significant ($p < 0.05$). One of the cases in Group 1 was passed away.
Results: Diagnosis of restrictive right ventricle physiology after the total correction of TOF has a negative effect on the morbidity and the mortality in early postoperative period. Early onset peritoneal dialysis in RRV + cases can prevent these negative effects.

P2194 - ENTERAL NUTRITIONAL THERAPY IN CHILDREN UNDERGOING EXTRACORPOREAL MEMBRANE OXYGENATION (ECMO) HOW TO DO IT

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Background: ECMO is a therapeutic modality that has been increasing in Pediatric Intensive Care Units (PICU). The enteral nutrition sometimes is delayed and many of these children remain fasting, due to the severity of the disease.
Objective: The objective of this study is to describe the enteral nutrition therapy in critically ill children treated with ECMO.
Methods: This is a retrospective study of consecutive children who underwent ECMO from November 2015 to December 2016; patient demographic, diagnosis, time of fasting, tube type, diet used, initial volume, time to reach full enteral nutrition, presence of pathological residues, vomiting, diarrhea and abdominal distension.
Results: Nine children were treated with venoarterial ($n = 8$) and venovenous ($n = 1$) ECMO in the period of the studying the PICU of a tertiary and private children's hospital in São Paulo, Brazil. The underlying conditions were acute respiratory distress syndrome, refractory septic shock, respiratory and circulatory failure after left pneumectomy, acute lymphoblastic leukemia and cardiogenic shock after Tetralogy of Fallot repair. The age ranged from 5 months to 7 years, of these, 5 (55,56%) received enteral nutrition by gastric tube starting 24 hours after being on ECMO, 3 (33,33%) stayed with parenteral nutrition due vomiting, diarrhea and abdominal distension and one child (11.11%) stayed in fasting due to side effects of chemotherapy, poor intestinal perfusion and intestinal bleeding. The enteral nutrition diet used was oligomeric (extensively hydrolyzed or elemental), without difference of tolerance between them, with initial volume of 20 mL/kg/day, in continuous or intermittent form, with an increment up to 50 mL/kg/day. Full enteral nutrition was achieved in up to 6 days
Conclusion: Although children on ECMO are critically ill, enteral nutritional therapy should always be established through algorithms that prioritize this nutritional modality.

P2209 - PEDIATRIC INFECTIVE ENDOCARDITIS DURING A PERIOD OF TEN YEARS IN A SINGLE COLOMBIAN INSTITUTION

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Background: Infective endocarditis (IE) is an entity with a growing incidence worldwide with an important morbidity and mortality. The aim of this study was to describe the epidemiological and clinical characteristics of the pediatric population with IE in a single institution over a period of ten years.
Materials and Methods: A descriptive retrospective study was performed. All pediatric IE cases (according to Duke criteria) between January 2002 and December 2012 were retrospectively reviewed. Ethics board approval was obtained and a waiver of consent was granted due to the retrospective nature of the review.
Results: 79 medical records were retrospectively analyzed, of whom 49 had a confirmed diagnosis of IE, according to Duke criteria. Most of the patients: 83.6% (41/49) had congenital heart defects (CHD), of which 63% (26/49) were acyanotic. IE was more frequent in patients with unrepaired CHD (36%, 15/49). 34% of patients (17/49) had a history of foreign material implantation, with patches, pacemakers and stents being the most frequent. Microbiological isolation was obtained in 76% (38/49) of the patients. The most frequently isolated germs were S. aureus (32%, 14/49) and S. coagulase negative (32%, 14/49). The most frequent anatomic localization was the tricuspid valve in 32% (14/49) of the cases, followed by the aortic valve with 30% (13/49). Mechanical complications were more frequent in patients with CHD ($p = 0.008$), while there were no significant difference in heart failure development ($p = 0.38$) or embolism ($p = 0.11$) between patients with and without CHD. 24% (12/49) of the patients required surgical management of the IE. The majority

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(57%) of the patients (28/49) had resolution of IE without sequelae, 20% (10/49) had any type of sequela and mortality was 18% (9/49).
Conclusions: This study contributes to a better knowledge of the clinical and epidemiologic characteristics of pediatric patients with IE in an important cardiovascular reference center in our country.

P2234 - EARLY POST OPERATIVE OUTCOME OF BIDIRECTIONAL GLENN SHUNT IN ADULTS

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Background: Nowadays, patients with univentriclar circulation and low pressure, low pulmonary blood flow are usually treated with a Bidirectional Glenn shunt (BDG). Operation is often in childhood and part of a staged palliation to the Fontan circulation. In developing countries many children remain untreated. However, a BDG may still play an important role in relieving symptoms in selected patients at adult age.
Methods and Patients: Retrospective study of all patients older than 16 years who underwent aBDG as palliative procedure for univentriclar circulation since 2009 in our center. There were 8 patients (4 male) with a median age of 21 years (17 to 28 years). Preoperative percutaneous oxygen saturation was 71% (65 to 78%) and hemoglobin 21 gm/dl (16.5 to 23.9 gm/dl). Mean pulmonary artery pressure was lower than 18 mmHg in all patients with normal size pulmonary arteries. Ventricular morphology was left ventricle in 4 patients, 3 patients had right ventricle morphology and one case with congenitally corrected transposition of great arteries with pulmonary obstruction.Two patients had a previous palliative arterial shunt and 2 patients had a pulmonary artery band. The remainder had native pulmonary stenosis.
Results: There were no operative deaths. Post-operative oxygen saturation on discharge was 83% (75-90%). Six patients were successfully extubated within 24 hours and the other 2 patients required mechanical ventilation for 48 hours. Two patients were re-intubated for another 24 hours due to type II respiratory failure and impaired ventricular systolic function, respectively. Inotropic support was needed in 5 patients for 48 hours. Two patients had a temporary supraventricular arrhythmia. There were no neurological complications or need for renal dialysis. Median hospital stay was 7 days (5-12 days).
Conclusion: BDG is a effective and safe palliative procedure in selected hypercyanotic adults with univentricular circulation.

P2284 - INFECTIVE ENDOCARDITIS IN CHILDREN EXPERIENCE OF A MOROCCAN CENTER

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Nicu, Mother and Child Hospital, Med Vi University Hospital, Marrakesh Medical School, Cadi Ayyad University, Pediatrics, Marrakesh-Morocco[1]; Pediatrics, Mother and Child Hospital, Med Vi University Hospital, Marrakesh Medical School, Cadi Ayyad University, Pediatrics, Marrakesh-Morocco[2]

Introduction: The impact of endocarditis in childhood accompanies many cardio-vascular disorders. Infective endocarditis (IE) is the most frequently occurring form of endocardium inflammation.
Objective: To study the epidemiological features of infective endocarditis.
Materials and Methods: In this retrospective study, we studied the clinical data, of all children with IE who had been admitted to the infection pediatrics department at the university hospital Mohamed VI Marrakech, from January 2011 to October 2016. The inclusion criteria were made based according to the modified Duke criteria.
Results: IE was found in 11 of 7627 hospitalized children, which represents a prevalence of 0.13%. The mean age was 5.1 years (Extremes [1 month; 15 years]), with a higher incidence in male (7 boys and 3 girls) and in children with cardiopathy (70%). In this study, based on the modified DUKE criteria: 7 cases were definite IE, and 3 cases were possible. Bi-antibiotherapy was the rule, of which 7 patients were treated with third-generation Cephalosporins-aminoglycosides, 2 patients with tri antibiotic therapy by adding amoxicillin and one case by rifampicin-teicoplanin. Evolution was favorable in 80% of cases and 20% had complications mainly neurological and thrombo-embolic, one of them died from pulmonary embolim.
Conclusion: Infective endocarditis occurs mostly in children with cardiac abnormalities. It is potentially lethal despite the progress made in the diagnosis, in the treatment with new effective antibiotic and surgery. IE remains a problem in our context in the absence of diagnosis of congenital heart disease, and lack of hygiene in certain urban area of the city.

P2290 - N TERMINAL PROHORMONE BRAIN NATRIURETIC PEPTIDE IN THE EVALUATION OF RESPONSE TO LEVOSIMENDAN IN PEDIATRIC PATIENTS. PRELIMINARY RESULTS

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Introduction: Besides common laboratory tests and echocardiographic assesment, an increasing number of biomarkers are proposed to evaluate treatment response and follow up in patients with cardiac failure.
Objective: To describe the usefulness of NT proBNP in the evaluation of response to Levosimendan (LEVO) in children with heart failure.
Design: Non interventional, observational and prospective study.
Setting: PCICU, Hospital "J.P.Garrahan", Buenos Aires, Argentina.
Patients and Methods: all patients with Levosimendan were included. Demographic data, cardiac diagnosis, surgical procedure, Rachs-1 score, mechanical ventilation, LOS and mortality were recorded, along with hemodynamic, and biochemical parameters. Every patient had a Pre LEVO echocardiographic evaluation performed by a senior cardiologist, NT-proBNP, lactic acid, urea and creatinine levels. All measurements were repeated after 24 hs, day 3 and 5 after LEVO. Statistical analysis: Hemodynamic, and biochemical parameters are analyzed before and after LEVO. Continuous variables are reported as median and interquartile range (IQR), categorical variables as absolute values or percentages. Wilcoxon Test was used to identify differences with $p < 0.05$ considered significant. (Stata 10.0).
Results: 13 patients were enrolled, the median age was 5 months (IQR 25-75: 2-15); median weight 4.95 kg (IQR 25-75: 2.8-7.5). 89% had complex procedures, RACHS greater than 3 most of them biventricular repair (83%). Median pre LEVO NT-proBNP

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was 7415 pg/ml (IQR 25-75: 4185-14001) and postLEVO at day 5 of 5896 pg/ml (IQR 25-75: 3851-7658) mz < (P 0.033). In 9 patients NT-proBNP showed more than 40% reduction between preLEVO and day 5 levels. Non statistical differences in echocardiographic measurements of ventricular function were found. Mortality rate is 7,69% (1 patient).
Conclusions; Preliminary data show a statistically significant decrease in NT-ProBNP values following Levo administration, with no echocardiographic differences in ventricular function.

P2306 - NEUROSURGICAL INTERVENTIONS IN CHILDREN WITH VENTRICULAR ASSIST DEVICES

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Background: There is limited published literature on the neurosurgical management of children who develop intracranial haemorrhage (ICH) while on Ventricular Assist Devices (VADs).
Objective: Describe patient demographics, management and outcome in children who required neurosurgical intervention on VADs.
Methods: Single centre retrospective observational cohort study of children (<16 years) on VADs. Outcomes assessed were survival at 30 days and 6 months.
Results: Out of 102 patients supported on VAD from November 2004 - September 2016, 5 developed ICH and required neurosurgical intervention in six instances (1 child had recurrent ICH). Three had haemorrhagic stroke [2 on Berlin heart Excor device (BH) and one on Heartware] and 3 subdural hematoma (2 on BH and 1 on Levitronix). ICH occurred between 17-103 support days on VAD. Infection concerns noted in 4 children (80%): fever, C - reactive protein (30-144 mg/L) with neutropenia in 2. Though no organisms were isolated, all were treated with empirical broad-spectrum antibiotics. One with recurrent ICH had inflammatory response - high CRP and fibrinogen with thrombus in VAD circuit. Wide fluctuations in activated partial thromboplastin time noted within 24 hours prior to ICH; anti-Xa levels remained in range. Neurosurgical interventions varied,with no immediate complications: burr hole, craniotomy and evacuation of ICH or external ventricular drain. Anticoagulation was withheld initially and unfractionated heparin restarted at low dose from day 4-8 post events. Four (80%) (including one with recurrent ICH) survived: 3 had heart transplant and 1 VAD explantation (myocardial-recovery). Neurologic outcome: residual hemiparesis in 3 children (one developed critical illness polyneuro-myopathy on nerve-conduction studies) and spastic-dystonic movement disorder in one child. All have subjective improvement in neurology during follow up.
Conclusion: Despite monitored anticoagulation, the bleeding risk in VAD patients increases with any underlying inflammation. We can safely perform neurosurgery on VAD patients with good short and medium term results.

P2343 - SPONTANEOUS CHYLOPERITONEUM AS A PREOPERATIVE COMPLICATION OF CONGENITAL HEART DISEASE

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Background/Hypothesis: Chyloperitoneum is a rare clinical condition, characterized by an accumulation of lymph fluid in the peritoneal cavity. There are a few reports of spontaneous chyloperitoneum secondary to congenital cardiopathies in the literature. We report a case of a spontaneous chylous ascites in a patient in the preoperative period.
Methods: case report and review of the scientific literature from published articles on the subject.
Results: Forty-days old Down Syndrome patient with Complete Atrioventricular Septal Defect (cAVSD), was hospitalized for decompensated heart failure. She presented with abdominal distension, and the ultrasound showed free liquid in the cavity. For anuria and edema, she got a Tenckhoff catheter for the peritoneal dialysis. Drainage of milky fluid was observed. Triglycerides of the peritoneal fluid was 133 mg/dL, characterizing spontaneous chyloperitoneum. Started fasting and total parenteral nutrition, and after 48 hours started minimal enteral nutrition with Monogen. Peritoneal liquid triglyceride control was 20 mg/dL. She underwent complete repair of the cAVSD, presenting bilateral chylothorax in the seventh postoperative day, and underwent vancomycin-based chemical pleurodesis on two occasions, with resolution of the condition. In addition, patient evolved with enterococcus sepsis and presumed fungal infection. During the investigation, secondary immunodeficiency was observed, attributed to the losses of immunoglobulins and lymphocytes due to the large volume of chylous liquid controlled with weekly infusion of Human Immunoglobulin.
Conclusions: Spontaneous chyloperitoneum associated with congenital heart defects or its clinical complications is still poorly described in the literature. We have a case where the spontaneous chyloperitoneum and the bilateral chylothorax were abundan,t leading to significant loss of immunoglobulins, leaving the patient susceptible to serious infections. Promptly treated with the specific therapies available, patient is now in good condition, in outpatient follow-up.

P2353 - CIRCULATING MICRO RNA208A AS A NOVEL EARLY BIOMARKER FOR THE POST OPERATIVE COURSE FOLLOWING CONGENITAL HEART SURGERY

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Background/Hypothesis: Clinical and biochemical markers used currently following pediatric cardiac surgery are poor predictors of the perioperative course. MicroRNAs (miRNAs), a class of short non-coding RNA molecules, have been shown to be promising biomarkers for several clinical conditions due to their tissue-specific expression pattern, rapid-release kinetics and stability in plasma. We hypothesized that cardiac-specific miRNA-208a, shown to be elevated in adults with acute myocardial infarction, can serve as an early, sensitive and specific circulating non-invasive biomarker for the post-operative clinical course in pediatric patients undergoing cardiac surgery.
Materials and Methods: RNA was extracted from serum samples obtained from 79 patients who underwent cardiac surgery pre-operatively and at 6, 12 and 24 h post-op. MiRNA-208a was quantified by quantitative PCR (QPCR). Correlations between the patient's course (surgical and clinical) and miRNA levels were tested.
Results: The amount of miRNA-208a rises sharply at 6 h after the operation (107-fold), remains high at 12 h (77-fold) and then

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declines. The amounts at 6 h and 12 h correlate significantly with the length of cardio-pulmonary bypass and aortic cross-clamp and with laboratory parameters as Troponin and Lactate. The increase in the amount of miRNA-208a at 12 h in patients with post-operative complications was 346-fold compared to 48-fold in patients without complications ($p < 0.05$). The amount of miRNA-208a at 12 h correlated with the length of hospitalization.
Conclusions: miRNA-208a is a suitable biomarker for predicting the post-operative course and complications in children following repair of congenital heart disease.

P2422 - SUCCESSFUL VENO ARTERIAL EXTRACORPOREAL MEMBRANE OXYGENATION IN CHILDREN WITH REFRACTORY SEPTIC SHOCK USING PERIPHERAL CANNULATION

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Background/Hypothesis: Children with refractory septic shock have high mortality. Extracorporeal membrane oxygenation (ECMO) is a possible treatment to these population. We aim to show positive outcome, achieving high flow rates, with VA ECMO using peripheral cannulation.
Materials and Methods: We have reviewed the hospital records for patients who needed ECMO between Aug/2015 and Dec/2016; patient demographic, diagnosis and outcomes were recorded.
Results: Four children with refractory septic shock were treated with VA ECMO during the study period. The mean age was 49,7 (13–104) months. The mean PIM II score was 28% (17%-37,7%), the mean vasoactive-inotrope score (VIS) was 235,8 (107,5-420). Before cannulation, the patients had pH 7,10 (6,99-7,22), lactate level 75,5 (18-122) mg/dl and PaO2/FiO2 ratio 117 (56-180). Micro-biological evidence of infection was present in 75%, streptococcus pyogenes was the most common agent. The mean time until cannulation after intubation was 15,7 h (10–20 hours). 3 children (75%) suffered cardiac arrest before cannulation requiring cardiopulmonary resuscitation (CPR). One of them was a cannulated during CPR (24 minutes). In these patients, ECMO was veno-arterial (v-a) with cephalad cannula. The maximum flow rate needed was between 100 and 150 ml/kg/min, achieved with no difficult with the peripheral cannulation. ECMO-related complications were unilateral vocal cord paralysis (50%), significant isquemic stroke (25%) and internal jugular vein stenosis (50%) and thrombosis (25%); All patients were place on Continuous Renal Replacement Therapy for fluid removal (50% continuous hemofiltration and 50% PRISMAFLEX). One patient also needed peritoneal dialysis. All patients were successfully off ECMO after 120 (62-184) hours (5 days). Three patients were successfully discharged from the hospital and one patient is still recovering, extubated.
Conclusion: ECMO using peripheral cannulation can be safely and effectively performed to rescue and support children with refractory septic shock.

P2427 - LRP1 AN ENDOCYTIC VESICLE TRAFFICKING PROTEIN ASSOCIATED WITH CONGENITAL HEART DISEASES AN UNEXPLORED PATHWAY

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Objectives: Congenital heart disease (CHD) remains the leading cause of neonatal deaths and affects 1% of live births per year in the United States. Strong evidence exists indicating a genetic role in CHD yet the specific mechanics and developmental processes are not fully understood. From a large-scale screen, we uncovered ten endocytic trafficking genes, including LRP1 (low density lipoprotein related receptor protein), that are associated with CHD. LRP1 regulates trafficking of internalized endocytic vesicles to different endosomal compartments, some destined for fusion with lysosomes and degradation, while others are recycled to the surface.
Methods and Results: Whole Exome sequencing from mutant line 1554 (MGI 96828) identified a missense (C4232R) mutation in the epidermal growth factor repeat domain of LRP1. Homozygote mutants exhibit an array of septation and outflow tract defects such as simple membranous ventricular septal defects (VSD), endocardial cushion defect/ atrioventricular septal defect, various types of double outlet of right ventricle (DORV) with normal great arteries or D-malposition of great arteries (Taussig-Bing anomaly). MEF (mouse embryonic fibroblast) derived from LRP1 $^{m/m}$ mutants demonstrate decreased expression of the protein at the cell surface and retention of the LRP1 mutant protein in the endocytic trafficking system. LRP1 is expressed in multiple cell lineages required for cardiac development. We use conditional gene ablation to generate several tissue-specific LRP1 mutants to identify the implication of LRP1 and its targets during endocardial and outflow tract development. LRP1-Nkx2.5-cre mice demonstrate various types of DORV (subaortic VSD, Taussig-Bing Anomaly), while LRP1-Tie2-cre (endothelium and endocardial cushion tissue deletions) demonstrate perimembraous VSD with 25% of mutants demonstrating AVSD. Second heart field specific LRP1 knockout mutants using Mef2C-AHF demonstrate simple VSD.
Conclusion: We provide evidence that endocytic trafficking plays an important role in CHD pathogenesis

P2436 - THE USE OF FAT FREE SEPARATED BREAST MILK AND PARTIAL (LIPID ONLY) PARENTERAL NUTRITION FOR POST OPERATIVE CHYLOTHORAX IN A RESOURCE LIMITED SETTING CASE REPORT TECHNIQUES AND LESSONS LEARNED

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Background: An 8 month 5 kg infant, Trisomy 21 post ASD/VSD/PDA closure developed 30mls yellow-opaque chest drainage following the first postoperative milk feed on postop day 0 (POD.0). ECHO revealed mild-moderate tricuspid regurgitation, mild pulmonary hypertension and dilated right ventricle. No formal analysis of drainage was undertaken. Whole milk feeding was stopped, drainage rapidly cleared and a combination feeding plan started. No medium chain triglyceride milk, oils, nor fat-free milk, were available.
Materials and Methods: Breast milk was collected and left standing, refrigerated and undisturbed for 24 hours, achieving natural and

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visible fat separation. Fat-free milk was aspirated from the base of the container, taking care not to aspirate the upper fat layer, approximately 4% by volume. Constant delivery was by syringe pump with vertical orientation so any residual fat accumulated at the syringe plunger was delivered. Juices and oral fruit purees were given to supplement breastmilk shortage Fat was delivered by Intravenous Lipid (LIPOFUNDIN MCT/LCT:10% BRAUN), peripherally commencing D1 at 0.5 g/kg/day, increasing to 3/g/kg/day D7. Fat soluble vitamin drops ADEK were added orally. Milrinone continued to POD.4, Oxygen POD.5. Sildenafil, Furosemide, Spironalactone throughout. Albumin 20% 1 g/kg/dose given per analysis.

Results: Maximal nutrition (POD.8) was calculated at 80-90 kcal/kg/day (50-60 kcal/kg/day enteral, 30 kcal/kg/day IV). Chest tubes, central venous line were removed POD.2, POD.3. Ultrasound revealed small bilateral pleural effusions on POD.3, undetectable by POD.5. A gradual re-introduction of full-fat breastmilk commenced on POD.10, was fully established on POD.12. No effusion was seen by ultrasound or x-ray following the reintroduction of enteral fat. Discharge home was achieved on POD14.

Conclusions: Human breast milk, with its nutritional and immunological advantages, can be safely utilised in postoperative chylothorax with no special equipment. Total parenteral nutrition (TPN) mandates central venous access with inherent infection risks. Peripheral venous delivery of lipid solutions is sustainable for many weeks and can be incorporated into any chylothorax strategy.

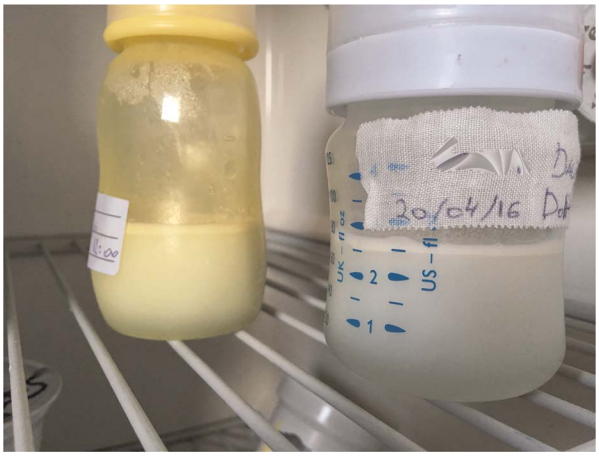

Figure 1.

Figure 2.

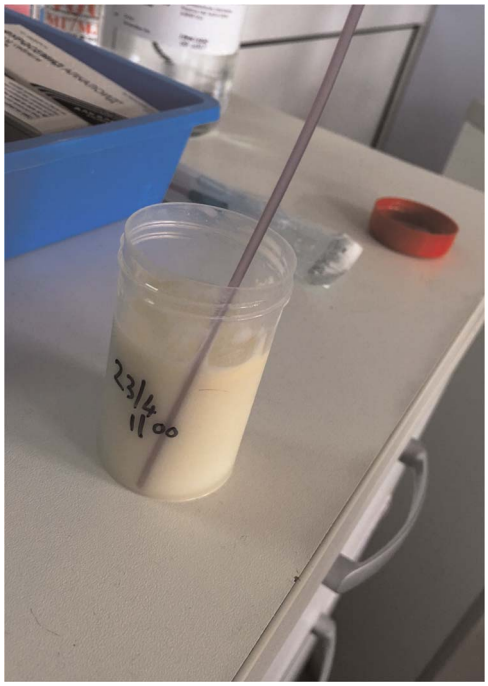

Figure 3.

P2438 - EXTRACORPOREAL LIFE SUPPORT IN A PEDIATRIC INTENSIVE CARE UNIT IN BRAZIL

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Background/Hypothesis: Pediatric patients with acute respiratory and/or circulatory failure may benefit from Extracorporeal Life Support (ECLS). The aim of the study is to describe the experience of a new ECLS program developed at the PICU of a private tertiary children's hospital in Brazil.

Materials and Methods: Retrospective study including all patients who had undergone non-postcardiotomy ECMO between July 2015 and December 2016; patient demographic, diagnosis and outcomes were recorded.

Results: Eight children were treated with ECLS. The mean age and weight was 33 months (5 months to 8 years) and 13,5 Kg (4,6-20 kg). Four children (50%) suffered cardiac arrest before cannulation requiring cardiopulmonary resuscitation (CPR). One of

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them was cannulated after 24 minutes of CPR (E-CPR). The underlying conditions were acute respiratory distress syndrome (n = 3), refractory septic shock (n = 4) and respiratory and circulatory failure after left pneumectomy (n = 1). Two children had newly diagnosed Acute Lymphoblastic Leukemia (ALL). All but one had peripheral cannulation with veno-arterial with cephalad cannula support. The other one had dual site venous-venous ECMO. ECMO-related complications were unilateral vocal cord paralysis (25%), significate isquemic stroke (12,5%) and internal jugular vein stenosis (50%) and thrombosis (25%); All patients were place on Continuous Renal Replacement Therapy for fluid removal (75% continuous hemofiltration and 25% PRISMAFLEX). One patient also needed peritoneal dialysis. Seven patients were successfully off ECLS after a mean of 216 (range 62 -621) hours (9 days). One patient died. Two patients are still recovering in the hospital, extubated. The other patients had hospital discharge.
Conclusions: In our center, the ECLS program has reached this far an 87,5% decannulation rate in non-postcardiotomy patients. A very committed multidisciplinary team and focus on continuous training may be the key feature to our successful program.

P2458 - CONGENITAL ABSENCE OF THE PERICARDIUM ASSOCIATED WITH ATRIAL SEPTAL DEFECT AND A DELETION OF CHROMOSOME 6Q13 IN YOUNG CHILD

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Congenital absence of the pericardium is an uncommon finding with variable presentations from asymptomatic to paroxysmal stabbing chest pain. Asymptomatic patients with congenital absence of the pericardium are typically discovered at an autopsy or during cardiac surgery. A 26-month-old female presented with severe dyspnea. Her past medical history and family history were unremarkable. A chest X-ray revealed levoposition of the heart into the left chest cavity, and severe pneumonia. An echocardiography demonstrated large sized secundum atrial septal defect with severe pulmonary hypertension. In cardiac tomography, we suspected a left lower pulmonary vein stenosis that comes from a compression of the aorta. We planned to perform a closure of atrial septal defect and a posterior aortopexy. Against our expectations, we discovered incidentally complete absence of the pericardium and no pulmonary vein stenosis during cardiac surgery, so we performed an anterior aortopexy, a fixation of pleura, and a closure of atrial septal defect. After surgery, she was improved from severe pulmonary hypertension and right ventricular hypertrophy. And we performed an array-CGH examination because of facial dysmorphism and developmental delay, and confirmed a gain of 2q21.1, and a deletion of 6q13, 6q14, 1q16.1. We experienced a case of congenital absence of the pericardium associated with atrial septal defect and a deletion of chromosome 6q13 in young child.

P2467 - RELATIONSHIP BETWEEN TROPONIN I VALUES AND POSTOPERATIVE OUTCOMES IN CONGENITAL HEART DISEASE CORRECTION REQUIRING EXTRACORPOREAL CIRCULATION

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Background and hypothesis: Troponin I levels are increased in the postoperative period due to myocardial damage. We proposed that the degree of elevation correlates with the incidence of renal failure and mortality.
Materials and Methods: Retrospective and prospective cohort study between November 2014 and December 2016, and included all children under 15 years of age who undergo to correction of congenital heart disease under cardiopulmonary bypass.
Results: A total of 144 patients were collected, with a median age of 3 years (9 days - 14 years), 63% with shunt heart disease, 8% with obstructive lesions and 29% with complex heart disease, the median time of perfusion was 87 Minutes (18-340 minutes) and aortic clamp of 39 minutes (0-116 minutes). The incidence of postoperative renal failure, we found a significant difference between those who did (percentile 50 of troponin I of 12.85 mcg/Lt (p25 of 5.77 mcg/Lt and p75 of 26 mcg/Lt)) in compared to the group without postoperative renal failure (troponin I 50th percentile of 3.97 mcg/Lt (p25 of 3 mcg/Lt and p75 of 9.33 mcg/Lt)). The mortality of our work was 12%, also finding a significant difference between patients who died (troponin I 50th percentile of 19.2 mcg/Lt (p25 of 10.3 mcg/Lt and p75 of 50 mcg/Lt)) and those that survive (troponin I 50th percentile of 5.5 mcg/Lt (p25 of 3.2 mcg/Lt and p75 of 15.5 mcg / Lt))
Conclusions: The levels of troponin I taken in the postoperative period of the correction of congenital heart diseases under extracorporeal circulation are statistically correlated with the incidence of renal failure and mortality.

P2491 - RISK FACTORS ASSOCIATED WITH MORBIDITY AND MORTALITY IN PATIENTS WITH HYPOPLASTIC LEFT HEART SYNDROME (HLHS) BROUGHT TO NORWOOD PROCEDURE AT A FOURTH LEVEL INSTITUTION IN COLOMBIA

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Introduction: Norwood procedure is the surgical option for patients with hypoplastic left heart syndrome (HLHS). In developing countries, the diagnosis of HLHS is usually late, access to specialized centers is limited, experience in the surgical approach is scarce and published reports are occasional with high mortality. The objective of this study was to analyze the preoperative, postoperative and surgical risk factors associated with morbidity-mortality and determine the survival in a 4th-level institution in a developing country.
Materials and Methods: Retrospective study. We reviewed the charts of patients with HLHS taken to Norwood procedure between January 2009 and December 2015. We analyzed preoperative, intraoperative and postoperative variables. Early mortality was measured (30-day and in-hospital); morbidity was determined by the use of ECMO, length of time on mechanical ventilation, inotropes, cardiac ICU, total days of hospitalization, neurological outcome and renal failure.
Results: Between January 2009 and December 2015, 55 patients diagnosed with HLHS had a Norwood procedure. 31 had modified Blalock-Taussig (MBT) shunt and 24 had right ventricle to pulmonary artery (RVPA) shunt. Mortality at 30 days was 23% and in hospital was 36%. Weight <3000gr (p 0.03) and mechanical ventilation (p 0.022) were the preoperative risk factors associated with death at 30 days. ECMO was a intraoperative and post-operative element related to both mortality (p 0.001).

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Non-restrictive atrial septal defect (p 0.011) combined circulatory arrest with periods of antegrade cerebral perfusion (p 0.024) and longer Cardiopulmonary bypass CPB (p <0.01) were risk factors for ECMO. Patients on ECMO had a higher incidence of renal failure (p 0.031). Arrest times and combined cerebral perfusion were associated with increased brain injury (p 0.04).
Conclusion: In our institution, risk factors associated with mortality are comparable with the literature. But contrary, the use RV-PA shunt was not a protective factor in the postoperative period.

P2510 - STUDY ON THE WARNING OF CENTRAL VENOUS OXYGEN SATURATION FOR CHILDREN WITH CONGENITAL HEART DISEASE

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Object To study the application value of ScvO2 in the post-operative care of children with congenital heart disease.
Methods: A retrospective analysis includes 201 cases as the research object from CICU of a tertiary children's Hospital in Shanghai, who after congenital heart disease surgery and accepted the dying notice. ScvO2 values detected when the child transfer into CICU and then every 12 hours until the child out of CICU or death. Analyze the difference between the ScvO2 values of the death group and the survival group, and the change of ScvO2 value.
Results: The maximum value of ScvO2, the lowest value, the mean interval is less than or equal to 50%, the mortality rate of children was 50% or above ($P < 0.01$). The maximum value of ScvO2 and the mean value is not less than 70.1%, the mortality rate was lower than 3.1% ($P < 0.01$). The minimum value of ScvO2 is more than or equal to 70.1% and the mortality rate is 0. After the first 12 hours of testing ,there was a rising trend and after 24 hours there was a significant downward trend until death. Survival group can be showed an upward or downward trend, in 24 hours it showed a rising trend and then tends to be stable.
Conclusion: ScvO2 monitoring is valuable for the care and treatment of the critically ill children with congenital heart disease.

P2514 - ALVEOLAR CAPILLARY DYSPLASIA WITH LEFT HEART OBSTRUCTION A CASE REPORT

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Case: We report a neonate delivered after 41 weeks of uncomplicated gestation, healthy family history, normal weight, APGAR, adaptation and early discharge after four hours. Eight hours later the newborn presented with severe hypoxemia (oxygen-saturation 55%) and dyspnea suffering from metabolic and respiratory acidosis (lactate 12 mmol/l). X-Ray appeared with enhanced vascular markings similar to respiratory distress syndrome. Echocardiography showed atrial septal defect type 1, aortic coarctation with hypoplastic arch, open ductus arteriosus with right to left shunt and consecutive volume depleted left ventricle. Despite immediate shock treatment by ventilation, inotropes, diuretics, and prostaglandin E1 low cardiac output syndrome and pulmonary hypertension persisted. Therefore implantation of extracorporeal membrane oxygenation (venoarterial ECMO) was necessary. Persistent pulmonary hypertension (PPHN) was addressed with nitrogen oxide, high frequency oscillation and surfactant admission. Nevertheless, neither lung function nore adequate systemic circulation could be established without ECMO. Having excluded other left heart obstruction causing pathologies and reasons for PPHN we performed open lung biopsy on ECMO. Histology revealed immature lobulae, thickened alveloar septae, medial hypertrophy of small pulmonary arteries and muscularization of distal arterioles - typical for alveolar capillary dysplasia (ACD). ECMO was explanted, therapy was switched to comfort care and the patient passed away on their parents arms at the age of ten days.
Conclusions: ACD is a rare and lethal lung disease associated with congenital heart defects and should be considered in obstructive left heart lesions and PPHN. As soon as other causes for left heart obstruction (intra-atrial membrane, pulmonary venous stenosis, APVC etc.) and PPHN (meconium aspiration, infection, congenital diaphragma hernia, pulmonary hypoplasia etc.) are excluded, lung biopsy is the option to define pathologies like ACD. FOX gene cluster examination can complete the diagnosis. Facing the fatal prognosis early, ante mortem diagnosis is essential to avoid futile interventions for patients, families and care providers.

P2553 - NEONATAL NON COMPACTED MYOCARDIOPATHY. PREDICTORS OF POOR OUTCOME

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Background: Non-compacted myocardiopathy (NCM) is a heterogenous myocardidal disorder characterized by abnormal trabeculations in the left ventricle. It is an increasingly recognized cause of cardiomyopathy in children and carries significant morbidity and mortality. Although much has been published in the last years, little is known about NCM in the neonatal period. The objective of this study is to characterize the involvement of newborns affected with NCM in two neonatal intensive care units (NICU) and to identify risk factors associated with increased mortality.
Materials and Methods: This is a retrospective study including all neonates diagnosed with NCM between 2006 and 2016 in the NICU. Diagnosis was based on echocardiographic findings. Data about prenatal history, gestational age and weight at birth, gender, age at diagnosis, left or biventricular involvement and associated malformations, medical and surgical treatments and evolution were collected.
Results: We identified fifteen patients diagnosed with NCM. Age at diagnosis was 7.2 ± 6 days. Twelve (80%) had isolated left ventricular involvement. Fourteen patients (93,3%) had other associated heart malformations; six had ventricular septum defects and six right side abnormalities. During admission, two patients presented arrhythmic events. During the follow-up period seven patients (46,6%) died. Patients who died had more biventricular involvement and more non-cardiac malformations (3/3[100%] vs 4/12[33%] $p = 0.02$). The main cause of death was ventricular dysfunction (85,7%). In the five cases in which the family authorized the autopsy the diagnosis was confirmed and two of them presented biventricular involvement not evidenced by echocardiography.
Conclusions: Neonatal NCM is associated with a poor prognosis. The higher mortality observed in our series (46,6%) is related not only to the high association with congenital heart disease, but also to a greater presence of early and severe left ventricular dysfunction. Poor ventricular function and biventricular involvement is a predictor of poor outcome.

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P2559 - ACUTE KIDNEY INJURY AFTER CONGENITAL HEART SURGERY FREQUENCY RISK FACTORS AND OUTCOME IN A DEVELOPING COUNTRY USING KDIGO DEFINITION

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Objectives: To determine the frequency, risk factors and outcome of acute kidney injury using KDIGO definition.
Methods and Materials: AKI was defined and staged as per KDIGO criteria using serum creatinine. Using this definition 150 patients who had AKI were matched to 75 patients who didn't had AKI from a total of 1680 consecutive patients between January 2007 to December 2016, making overall incidence of AKI to be 8.9% in our series. Appropriate statistical tests were applied to determine the association of different variables with development of AKI and P value of <0.05 was taken as significant.
Results: Both the groups were similar in age (mean age 4.5 years and 6 years), gender (64% male in cases and 60% males in controls) and pre op creatinine (0.35 mg/dl in cases and 0.46 in controls), RACHS scoring and anatomy. Increased cardiopulmonary bypass time and aortic cross clamp time, postoperative hypotension, postoperative inotropic support, need for blood transfusion, use of nephrotoxic drugs, low cardiac output syndrome, development of postoperative sepsis were significantly associated with development of AKI on univariate and multivariate analysis. 40 patients had stage I AKI, 60 had stage II and 50 had stage III AKI. 10 patients required peritoneal dialysis, mean length of stay was 5 days in cases and 3 days in controls, 21 patients in cases and 1 patients in controls expired during the study period. Data collection and analysis is still in progress.
Conclusions: AKI is a very common problem in postoperative congenital heart surgery patients associated with higher mortality and morbidity.

P2604 - VENO ARTERIAL EXTRACORPOREAL MEMBRANE OXYGENATION FOR ACUTE VIRAL MYOCARDITIS WITH CARDIORESPIRATORY ARREST IS IT TOO LATE TO START

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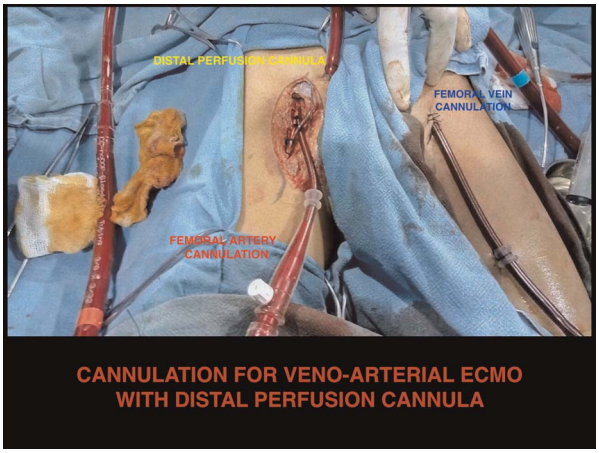
Background: Severe ventricular dysfunction is one of the dreadful complication of viral myocarditis. We are sharing our experience of three cases presented to tertiary cardiac care unit with cardiorespiratory failure and arrest, subsequently needed ECMO support.
Methods, Case Details and Results: Between October 2015 to October 2016, three patients presented with viral myocarditis, severe LV dysfunction, bradycardia and hypotension on arrival to emergency room. Veno-arterial ECMO (femoral vein and artery in 2, internal jugular vein and carotid artery in one) was initiated in all while continuing or immediately after Cardio pulmonary resuscitation (CPR). Median weight and age were 22 kg (range 9.7-64 Kg) and 8 years (range 11/2-13 Years) respectively. Median duration of CPR was 90 minutes (range 60 - 100 minutes). Median Duration of ECMO was 6 days. Viral profile were send for all patients but only one showed positive virology for Echovirus and Coxsackie virus. Two patients requires CRRT while on ECMO due to acute renal failure. Two out of three patients having good LV mass index to begin with were successfully weaned off ECMO with good cardiac recovery. 8 year old girl for which virology was positive got discharged without any neurological deficit. On follow up she showed good ventricular ejection fraction and symptom free status but developed loss of right leg distal pulses without sign of limb ischemia. 11/2 years old weaned off successfully from ECMO but died after 3 days due to DIC and septic shock while for 13 year old ECMO support was withdrawn in view of cerebral uncal herniation.
Conclusion: ECMO can be initiated even during or immediately after CPR with reasonable outcome. Effective CPR and prevention of multiorgan dysfunction are key for successful outcome. Therefore ECMO should be considered before onset of end organ damage.



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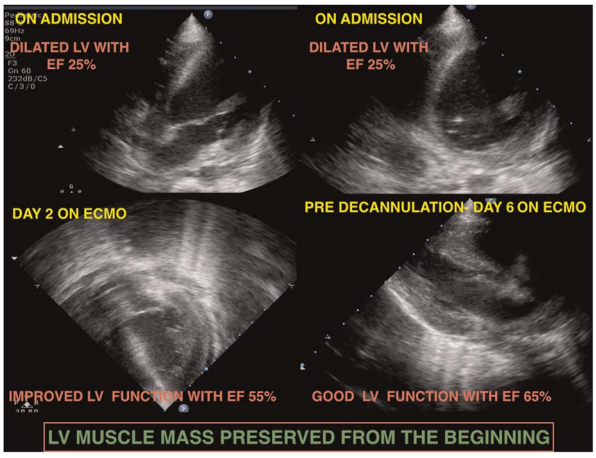

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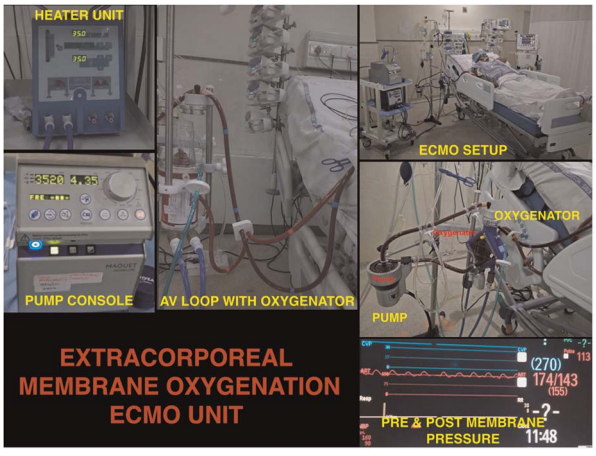

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P2663 - CRITICAL CONGENITAL HEART DISEASE SINGLE INSTITUTIONAL EXPERIENCE AND OUTCOMES IN A LOW MEDIUM INCOME COUNTRY

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Background: Critical congenital heart disease is associated with high morbidity and mortality. The factors associated with increased neonatal morbidity and mortality include prematurity ≤35 weeks, low birth weight, hypoplastic left heart syndrome and complex cardiac anatomy. major extracardiac and genetic malformations, neonatal sepsis and meconium aspiration.Early diagnosis and intervention can be achieved with prenatal diagnosis and pulse oximetry screening.
Methods: A retrospective review of cases admitted in the Neonatal Intensive Care Unit (NICU) and High Care between 2005 and 2015 and presenting to the Division of Paediatric Cardiology at the Chris Hani Baragwanath Academic Hospital.There are limited prenatal screening services, no onsite cardiothoracic surgical and pulse oximetry screening services.Data related to demographics, prenatal and postnatal diagnosis,clinical presentation,treatment, surgery,mortality and follow up was collected.
Results: The diagnosis of critical congenital heart disease was made in one hundred and twenty patients (males, 62.7%). Median age at diagnosis = 2 days (1-78 days, IQR = 1-5). Mean age at presentation = 2.1 ± 2.8 days (1-18 days). Twenty three cases (19,2%) were preterm ≤35 weeks of gestation.Fifty two cases(43%) had low birth weight, mean birth weight = 1.91 ± 0.39 kg (0.97-2.49 kg). Prenatal diagnosis was made in nine patients (7.5%). Most common presentation was respiratory distress, cyanosis and shock with metabolic acidosis. Majority required invasive positive pressure ventilation (IPPV) and sixty nine patients (57.5%) received PGE2 infusion. Common cardiac lesions were hypoplastic left heart syndrome, transposition of the great vessels and pulmonary atresia with or without ventricular septal defect. Extracardiac and genetic malformations were diagnosed in sixteen patients (13.3%). Catheter interventions were done in eighteen patients (15%). Surgery either palliative or corrective was achieved in twenty two patients (18.3%). Forty five patients (37.5%) were deemed inoperable. Overall preoperative mortality was 71,7%. Preoperative mortality without the patients deemed inoperable was 34.2%.
Conclusions: Mortality is high in patients with critical congenital heart disease especially in those presenting during the neonatal period.Inadequate prenatal screening, prematurity, low birth weight,extracardiac anomalies, lack of pulse oximetry screening program and resources may have contributed to the mortality.

P2697 - ADVERSE EVENTS REPORTING A PROGRAM TO IMPROVE QUALITY AND SAFETY IN A PEDIATRIC CARDIAC INTENSIVE CARE UNIT IN ARGENTINA

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Objective: To describe the implementation of an adverse event reporting program in a pediatric cardiac intensive care unit (PCICU).
Setting: PCICU. Garrahan Hospital, Buenos Aires, Argentina. Material and Methods: a multidisciplinary group was established to develop the program. A special form to collect selected events was designed. Events were grouped as safety (S), medication (ME), mechanical ventilation (MV), vascular lines (VL), procedures (P), nutritional support (NS), surgical (Q) and in-hospital transfer (T). All staff were trained about the program goals and how to complete the forms. A mailbox was placed in a visible place inside the unit. Any team member can report events, is voluntary and anonymous. Monthly reports are analyzed and preventive measures and/or process changes are proposed.
Preliminary Results: There were 178 events reported between December 2015 and December 2016. The most frequently reported were30% vascular lines, 24% medication,16% Safety and 8% mechanical ventilation. More than 85% were reported by nurses. 33% in the morning shift and 30% during the night one. Increasing bed side strategies and communication improvement were proposed as a result of this analysis.
Conclusions: Nurses participation in the program was very high. Adverse events reporting is a new tool which provides valuable information to provide safer and higher quality carein our PCICU.

P2715 - HEALTH PROFESSIONALS INTENSIVE CARE UNITS AND DEATH WHAT IS THE RESULT OF THIS EQUATION

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Background: Death is constantly present in the daily processes of intensive care units (ICUs), and it has different implications for health professionals.
Objective: To analyze factors associated with the meaning of death in the ICU and the attitudes of health professionals who are faced with death.
Design: This cross-sectional study considered 5 different outcomes: "meaning of death"; "meaning of death of terminally ill patients"; "attitude towards giving news of death"; "resuscitation of a patient without possibility of treatment"; and "discussion about do-not-resuscitate orders". The explanatory variables were demographic, socio-cultural and vocational training characteristics.
Setting/Subjects: Doctors and nurses in the ICUs (general, cardiac, pediatric and neonatal) of a university hospital in northeastern Brazil.
Measurements: Associations were estimated using a multiple regression analysis ($\alpha = 5\%$).
Results: A total of 52 doctors and 45 nurses were included. The following were associated with the meaning of death for ICU professionals: personal experience with death, professional category, type of ICU and length of service ($P < 0.05$). The attitudes of these professionals when facing death were associated with the following: importance attached to religion, professional category, length of training, discussion of the topic in the professional life, the patient's age in the first professional experience with death and the number of units in which they had worked ($P < 0.05$).
Conclusions: The death of ICU patients is associated with pain, guilt and failure for many doctors and nurses, and their attitudes reveal a strong therapeutic obstinacy. Perceptions and attitudes towards death are affected by socio-cultural aspects, previous experience and professional training. Curricular changes and

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changes to the work process of these professionals are suggested, including the incorporation of protocols for palliative care and the communication of bad news.

P2787 - GASTROSTOMY IN INFANTS AFTER CARDIAC SURGEY INCEDINCE AND ASSICIATED RISK FACTORS

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Background/Hypothesis: Feeding difficulty following congenital heart surgery in the neonatal period is a known complication. The aim of this study is to identify the risk factors most associated with gastrostomy placement.
Materials and Methods: We conducted a retrospective chart review of patients 0-3 months of age with congenital heart surgery and gastrostomy placement with and without fundoplication from our institutional Pediatric Research Database from July 2010 - July 2015. 37 patients were identified and 29 included in the study. A control group of 123 patients was identified and used for analysis. Preoperative, intraoperative, and postoperative factors of interest were collected. Univariate regression and multivariable logistic regression analysis of these factors was performed to identify those associated with gastrostomy placement in our cohort.
Results: In the full cohort of 152 patients, 37 patients underwent gastrostomy placement (24%) after surgery for congenital heart diseases. 8 patients were excluded for different reasons, 29 were included. The main two factors were found to be associated with need for Gastrostomy placement are vocal cord dysfunction (VCD) (odds ratio 7.0; 95% confidence interval, 1.8-27.2; $P = 0.004$) and need for surgical revision (odds ratio 7.9; 95% confidence interval, 2.5-25.4; $P = < 0.001$). Other risk factors like small body weight, high RACHS score, presence of genetic anomaly, and the need for prolonged Total parental nutrition or promotility agents were statistically significant using univariate analysis but not using multivariable regression analysis.
Conclusions: In infants undergoing congenital heart surgery in the first three months of life, patients with VCD or need for surgical revision are at a higher risk for gastrostomy placement. These results indicate a need for the early evaluation and reduction of injury leading to VCD and surgical revision.

P2816 - MULTIPLE LUNG ABSCESS SECONDARY TO SEPTIC EMBOLISM DUE TO INFECTIVE ENDOCARDITIS. PEDIATRIC CASE REPORT

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Background: Septic pulmonary embolism is a serious and unusual condition where the focus is primarily the right infective endocarditis (IE) and less frequent from thrombophlebitis or osteomyelitis. The most cases reports of pulmonary embolism secondary to IE affect intravenous drug users or immunocompromised adults patients.
Method: Case of an inpatient at Cardiac Critical Care Unit at Children's Hospital Roberto del Rio in Santiago de Chile in October 2016 is reported.
Results: We described a 2 years old female, with history of small ventricular septal defect (VSD), consulted for 15 days of fever, illness appearance, change in cardiac murmur, with elevated C protein reactive, leukocytosis and thrombocytopenia. Color Doppler echocardiography demonstrated a small perimembranous VSD and a vegetation in septal tricuspid leaflet. Treatment with ceftriaxone and amikacin was initiated. Blood cultures for Streptococcus viridans were positive. Due to no response to antibiotherapy and onset of cough, at fifth day of treatment heart surgery was performed, vegetation was removed and VSD closure, without complications. The patient evolved with persistent fever, right interstitial infiltrate at chest radiograph (CR). Computed tomography angiography (CT angio) of chest images showed signs of pulmonary embolism with multiple areas of pulmonary infarction perfused by right pulmonary artery, thereby at ninth day of treatment, subcutaneous enoxaparin was started. Due to clinical condition and CR evolves with air-fluid levels, the antibiotherapy was changed to vancomycin with meropenem. New CT angio confirms a large air-fluid collection that compromises the right lower lobe. Percutaneous drainage was performed and collection cultures were positive to Pseudomonas aeruginosa carbapenem resistant and sensitive to ceftazidime and amikacin for which treatment was changed, achieving patient recovery.
Conclusions: In pediatrics patient the septic pulmonary embolism represent an exceptional situation. We report this case to mention the importance of basic clinical parameters and the relevance of early diagnostic recognition.

P2821 - NON WHITE RACE IS A RISK FACTOR FOR MORTALITY IN PEDIATRIC CARDIOMYOPATHY AND MYOCARDITIS PATIENTS

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Background: Previous studies identify non-white race as a risk factor for mortality in patients undergoing extracorporeal membrane oxygenation (ECMO) or cardiac surgery. We investigated the association between race/ethnicity and outcomes in pediatric patients with cardiomyopathy and myocarditis using a national database.
Methods: All pediatric patients with a diagnosis of myocarditis or cardiomyopathy and without congenital heart disease in the Healthcare Cost and Utilization Project Kids' Inpatient Database from 2003 to 2012 were included. Logistic regression models were created to explore race and mortality, adjusting for year, sex, age group, diagnosis, insurance, hospital type, and organ dysfunction.
Results: A total of 34,617 patients with cardiomyopathy (71%), myocarditis (25%), or both (4%) were included. The identified racial/ethnic groups were white (39%), black (20%), Hispanic (15%), and "other" including Asian/Pacific Islander, and Native American (8%). Black race (Odds Ratio (OR): 1.28, 95% Confidence Interval (CI): 1.04-1.58) and Hispanic ethnicity (OR: 1.30, 95% CI: 1.05-1.62) were independent risk factors for mortality after adjusting for the above covariates. Black race was also a risk factor for cardiac arrest (OR: 1.25, 95% CI: 1.03-1.51) and ECMO usage (OR: 1.55, 95% CI: 1.07-2.25). After adjustment for mechanical circulatory support (ECMO and ventricular assist devices) and heart transplantation, black race was no longer associated with mortality. However, Hispanic ethnicity remained associated with poor outcome (OR: 1.31, 95% CI: 1.04-1.64).

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Conclusion: Black race and Hispanic ethnicity are risk factors for mortality in pediatric patients with cardiomyopathy or myocarditis. In black patients, this may be due to disparities in supportive therapies.

P2822 - CAN NEWBORN WITH CONGENITAL COMPLEX HEART DISEASE BE BORN BY NORMAL DELIVERY

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Introduction: Prenatal diagnosis of complex congenital heart disease is frequently diagnosed and usually perinatal team propose a planned cesarean section. However, the benefit of this behavior is not yet clear.
Objectives: To determine if infants with prenatal diagnosis of complex congenital heart disease had better Apgar test than those without diagnosis or those born with normal delivery.
Patients and Methods: Retrospective study between January, 2014 and August, 2016, review of records of newborns with diagnosis of postnatal congenital heart disease. Complex congenital heart disease was defined as those that requiring some type of surgical intervention during the first year of life.
Results: Twenty-nine patients with complex congenital heart disease from a total of 11,275 live births (2.57 × 1000 NV, 16 per spontaneous delivery (56%) and 13 cesarean (44%) were born, 20% were emergency cesarean sections. Median APGAR of the complete group at 1 minute and at 5 minutes was 8 and 9, respectively. For the spontaneous delivery group, it was 8.5 and 9. For the cesarean group it was 8 and 9. The most frequent causes of cesarean section were: previous double cesarean section (50%), maternal cause (20%) and dystocia (15%). The prenatal diagnosis of congenital heart disease reached 52% and the most frequently diagnosed cases were left ventricular hypoplasia (n: 6) and atrioventricular septal defects(n: 6). Only 50% of these patients had prenatal diagnosis and half of them were born by normal delivery.
Conclusions: Diagnose of complex congenital heart disease does not imply a programmed or cesarean delivery, but it would be reserved for situations in which the maternal condition warrants it.

P2827 - IMPACT OF LEVOSIMENDAN IN HEART FAILURE PEDIATRIC PATIENTS

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Background: Levosimendan is an inodilator agent used in the treatment of low cardiac output heart failure in pediatric patients. We present a single center's experience regarding the use of levosimendan in children, evaluating its role in cases of ventricular dysfunction and low cardiac output syndrome.
Material and Methods: Patients were selected for levosimendan administration if low cardiac output heart failure was present either because of dilated cardiomyopathy or ventricular dysfunction after cardiac surgery. Retrospective analysis of data of all children treated with levosimendan from 2004 to 2016. Each patient received at least one 0.05-0.2 μg/Kg/min infusion of levosimendan, without a loading dose. Biomarkers and echocardiographic data before and after treatment were compared.
Results: Twenty-nine patients were included. The median of age was 2 years (2 months-19 years) and 65.5% of patients were female. More than 50% of all patients had dilated cardiomyopathy. Thirty-one percent had ventricular dysfunction after cardiac surgery. Levosimendan improved lactate levels (11.4 to 7.4 mmol/L, $p = 0.008$, $d = 0.85$), glomerular filtration rate (97 to 148 mL/min/1.73 m2, $p < 0.001$, $d = 1.22$), B type natriuretic peptide concentration (2655 to 498 pg/mL, $p = 0.001$, $d = 1.36$) and left ventricular fractional shortening (16.9 to 22.7%, $p < 0.001$, $d = 1.53$). After treatment, there was a trend towards left ventricular (LV) dimension reduction although it did not reach statistical significant difference (LV z-score 4.7 to 4, $p = 0.102$). There were no complications during the treatment.
Conclusions: Our results showed cardiac and renal function improvement with a better hemodynamic and metabolic profile. Although there is still little evidence about the use of levosimendan in children, these findings are consistent with other reported studies.

P2854 - MILRINONE AND ARRHYTHMIA FOLLOWING PEDIATRIC HEART SURGERY IS THERE AN ASSOCIATION

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Hospital Santa Cruz - Centro Hospitalar Lisboa Ocidental, Pediatric Cardiology, Lisbon-Portugal[1]; Hospital Santa Cruz - Centro Hospitalar Lisboa Ocidental, Cardiothoracic Surgery, Lisbon-Portugal[2]

Background: Milrinone is an inotrope and vasodilator used after pediatric heart surgery. Arrhythmia is considered to be one of its most frequent side effects, but with scarse evidence in pediatric age. In our study, we evaluated the incidence of arrhythmia and its potential association to milrinone following pediatric heart surgery.
Materials and Methods: We performed a retrospective study in a Pediatric Cardiac Intensive Care Unit. Patients with ≤1 year of age submitted to heart surgery with extracorporeal circulation during the last 4 years were randomly selected. The data was analyzed by SPSS, using the Chi-square or Fisher's exact test for categorical variables and the T-test for ordinal variables.
Results: Our sample consisted of 104 patients, median of age 4 months. Diagnosis included: transposition of the great arteries (29), ventricular septal defect (25), tetralogy of Fallot (18), complete atrioventricular septal defect (14), others (25). Arrhythmia was documented in 36.5% of the patients (38): junctional tachycardia (19), atrioventricular block (5), ventricular extrasystole (5), others (9). The following factors were significantly associated with arrhythmia: coronary manipulation ($p = 0.019$), RACHS1 ≥ 4 ($p = 0.017$), use of milrinone ($p = 0.003$) or epinephrine ($p = 0.008$), and with lower significance, with the occurrence of low cardiac output syndrome ($p = 0.041$) or acute kidney injury ($p = 0.028$). Patients with arrhythmia were significantly younger (3.1 vs 5.4 months, $p = 0.004$) and presented higher lactate levels (5.4 vs 3.0 mmol/L, $p = 0.011$).
Conclusion: In our study we found a statistical association between the occurrence of arrhythmia and the use of milrinone after pediatric heart surgery. This may result from a pure side effect of milrinone or a cluster of factors that configure situations of higher arrhythmia risk.

P2900 - IDIOPATHIC ARTERIAL CALCIFICATION IN NEONATE WITH HYDROPS FETALIS – DIAGNOSIS AND THERAPEUTIC MANAGEMENT

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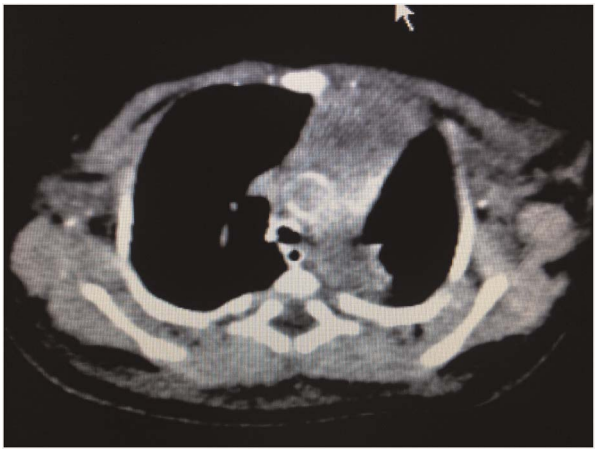
Introduction: Idiopathic arterial calcification (IAC) is a rare and generally fatal autosomal recessive disorder of unknown etiology. IAC is characterized by extensive and diffuse calcification and stenosis of the medium and great arteries due to deposition of calcium in the internal elastic lamella, which culminates in heart failure and recalcitrant arterial hypertension.
Case Report: Woman at 35 weeks of gestational age was referred to the tertiary hospital with diagnosis of hydrops fetalis and myocarditis. Newborn with 3,840 g, male, resuscitation was performed. He presented anasarca, distended abdomen, hepatosplenomegaly, hypophonetic heart sounds, cardiomegaly, arterial hypotension and anemia. Echocardiography revealed significant pericardial effusion, left ventricular dysfunction, and hyperechogenic arterial walls. IAC was hypothesized and later confirmed by angiotomography, which demonstrated diffuse arterial involvement (aorta; iliac, renal, femoral arteries, pulmonary artery). The newborn was submitted to pericardiocentesis, which improved the hemodynamic parameters. Treatment with sodium pamidronate was initiated on the fourth day after birth and lasted three days. The newborn improved progressively and was extubated at 10 days of age.The newborn needed amlodipine due systemic arterial hypertension. Sodium pamidronate was administered every two months for 12 months and every six months for further 12 months, until he was aged 24 months. The patient is currently 29 months old and stable. He has satisfactorily gained height and weight and new medication cycles are being planned. The latest echocardiogram has shown improved function of the left ventricle. According to the most recent angiotomography scan, arterial calcification has decreased significantly.
Comments: Although IAC is a rare condition with highly morbimortality, it should always be investigated in fetuses or newborns with arterial parietal hyperechogenicity and hydrops. In this case report, early echocardiographic findings combined with specific support therapy (sodium pamidronate) seem to have been crucial factors in the patient's initial progress. Serial follow up should be accomplished.



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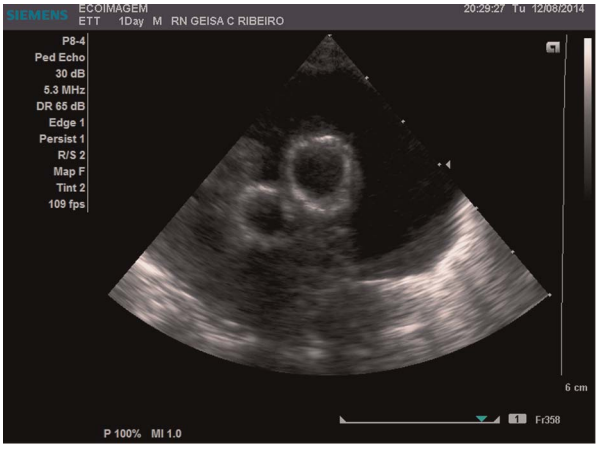

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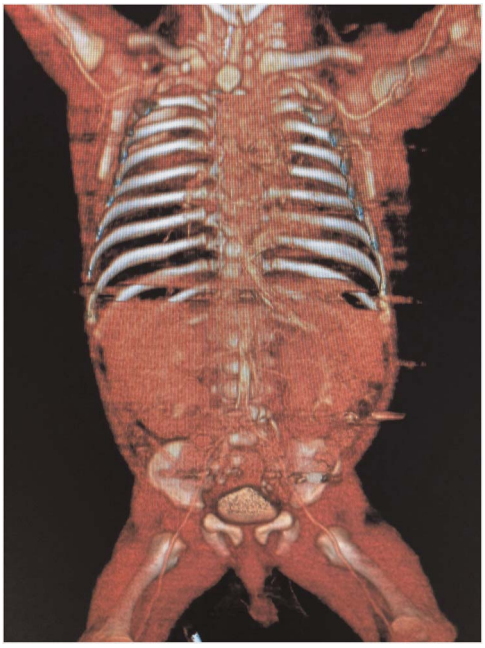

Figure 3.

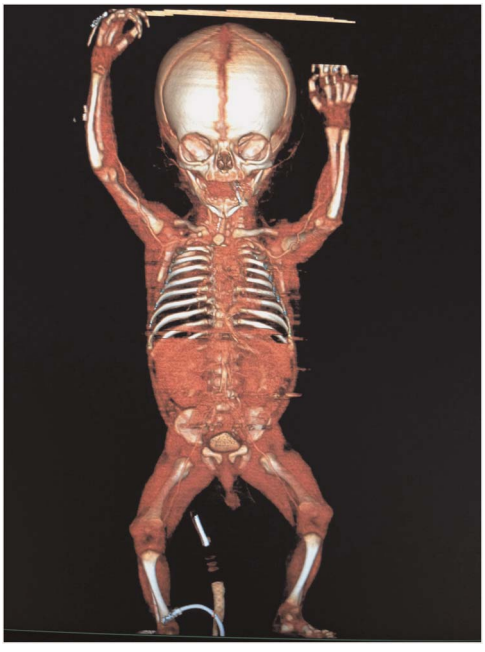

Figure 4.

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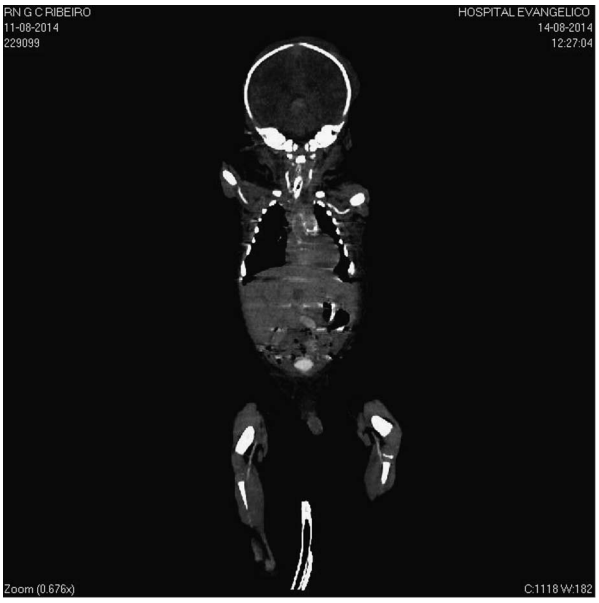
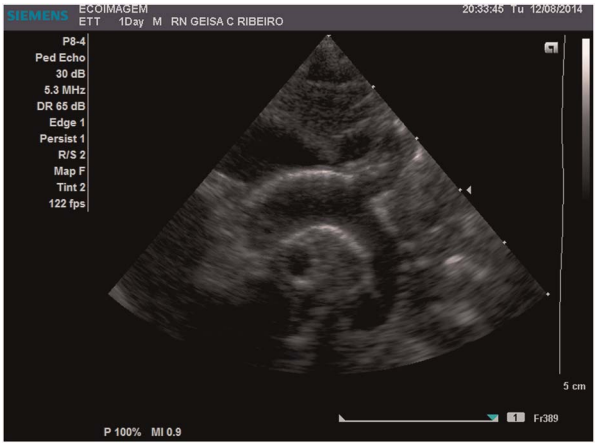


Figure 5.



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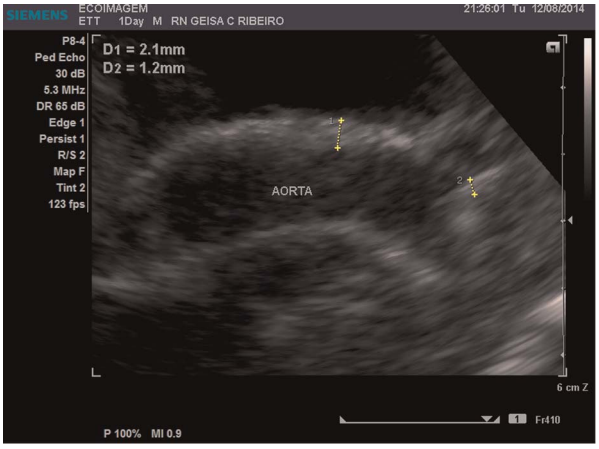

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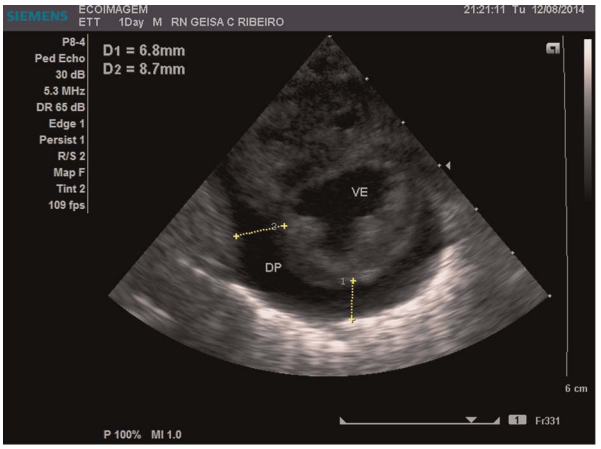

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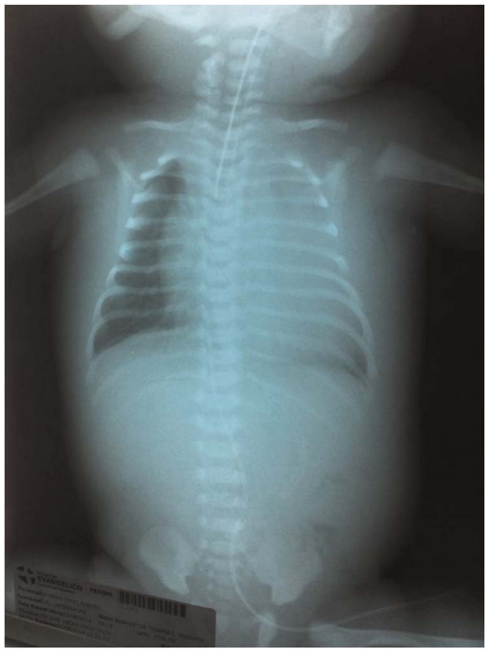

Figure 9.

P2928 - VENOUS TO ARTERIAL CARBON DIOXIDE DIFFERENCE A MARKER OF LOW CARDIAC OUTPUT IN CHILDREN AFTER CARDIOVASCULAR SURGERY

Mercedes Montonati[1], *Jessica Claus*[2], *Guillermo Moreno*[1], *Cecilia Quaglia*[2], *Maria Balestrini*[2], *Ricardo Magliola*[1]
Garrahan Hospital, PCICU, Capital Federal-Argentina[1]; *Clinic Y Maternity Suizo Argentina, PCICU, Capital Federal-Argentina*[2]

Objective: The purpose of this study was to correlate central venous to arterial carbon dioxide difference (PCO2 gap) with mortality as the primary end point and low cardiac output syndrome (LCOS) and their relationship to other markers of impaired tissue perfusion in patients after cardiac surgery with bypass (CPB), and the positive predictive value.
Design: prospective, observational clinical study. Setting: PCICU. Material and Methods: All patients less than 6 months of age that were admitted to the PCICU after CPB. Measaurements of

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arterial and venous CO2 were performed in every patient at 6, 16 and 24 hrs. Values of PCO2 were calculated by subtracting venous CO2 from arterial CO2. For the purpose of the study, CO2 gap >6 and ScvO2 < 70% represented hypoperfusion. Other parameters of global blood flow were assessed, venous mixed saturation (VMS), lactate (AL), NIRS, urine output and vasoactive inotropic score (VIS). We defined LCOS with a VIS >15. Data was expressed as mean and SD or percentage. Chi square test for categorical variables and spearman analysis to measure the degree of correlation between the variables.
Results: 30 infants were enrolled from 8/2016 to 1/2017. 40% were neonates, 60% male. The median weight was 3,1 kg (r2,7-8,8), more 50% were RACHS >3 and 40% had VIS > 15. In relation to the PCO2 gap, 60% of the patients with PCO2 gap >6 had ScvO2 >70%. We also finded that at 12 hrs. postoperative (PO) we had a PCO2 gap >6 with a lactate value >20 in all our patients.
Conclusion: All patients had LCOS. The widening of the CO2 gap poorly correlates with ScvO2.The first 12 hrs. postoperative we have better correlation between CO2 gap and LA.

P2932 - USE OF VD VT (DEATH VOLUME TIDAL VOLUME) AND CO2 EXHALED VOLUMETRIC AS REGARDS CLINICAL PARAMETERS LIKE PREDICTORS OF EXTUBATION IN POSTOPERATIVE PATIENTS OF CARDIAC CONGENITAL DISEASE

Jessica Claus[1], Cecilia Quaglia[2], Leonardo Perez[1], Alicia Saenz[1], Lucrecia Braco[1], Viviana Miranda[1], Gabriel Arias Miño[1], Maria Balestrini[1]
Clinic and Maternity Suizo Argentina, Pcicu, Capital Federal-Argentina[1]; Clinic Y Maternitand Suizo Argentina, Pcicu, Capital Federal-Argentina[2]

Objective: Analyze the value of Vd/Vt and CO2 exhaled volumetric like predictor of the success in the extubation in the PCICU.
Material and Method: Study of cohort, prospective, longitudinal and observational in patients <1 year from 1/2016 to 1/2017 with bypass (CPB) in assistance of mechanic ventilation (VM), which extubation criteria. We valued age, weigh, cardiac congenital disease with categorization of RACH1, vasoactive of inotrópics score (VIS), parameters VM: paFio2 (Pafi), Index of oxygenation (OI). Lactic acid (LA), CO2 volumentrics and interrelation with PaCO2, and Vd/Vt measured at 0,6,12 and 24 hrs. And a Vd/Vt relation <0,4 might relate successfully in the extubation and a relation >0,65 to defeat.
Results: 30 patients, with CPB, with medium 3,5 month and weight of 3,8 Kg, RACHS 1: 1 2% ; 2 43%, 3 48% and 4 7% VM: PaFi median 435, OI: median 2, VIS was 40% < 15, LA at the moment of the extubacion <20, The measurement of CO2 volumetric pre-extubacion 32,5 in interrelation with a Vd/Vt <0,5 when they arrived, the first day <0,5 and at the moment of extubation <0,5. Of 30 patients 5 went to nasal cannula with good tolerance without request of non invasive positive pressure (NIPP) and 25 went to cannula but they needed of NIPP a few minutes after the extubation. The analyzed patients presented at the moment of being extubated a Vd/Vt <0,6, nevertheless considering the characteristics of this group of patients the majority needed the use of NIPP. However patients did not need reintubation to VM.
Conclusion: The value of Vd/Vt contributes an important fact at the moment of the extubation, therefore we can deduce that it is a good factor predictive for successful extubation; although we cannot predict if the patient is going

P2937 - DIFFERENCE OF RESUSCITATION OUTCOME BY THE CAUSE OF SUDDEN CARDIAC ARREST OF SCHOOL STUDENTS

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Nihon University, Pediatrics and Child Health, Tokyo-Japan

Background: Basic Life Support and Automated External Defibrillator have been contributing to decrease sudden cardiac death in school since their introductions. It is necessary to evaluate the latest results of resuscitation according to the cause of cardiac arrest.
Methods: We analyzed all reports including ECG, death or health certificates, and autopsy reports of all decease and recovered cases registered to the mutual aid system for schools between 2008 and 2013.
Results: There were 121 deceased cases and 207 resuscitated cases during these 6 years. Incidence (/100,000studente·year) of decease and recovery were 0.12 and 0.20, respectively. Estimated cause of SCA was unknown in 28% and 43% ($p < 0.01$) of decease and recovered cases, respectively. VF ($p = 0.58$), LQT ($p = 0.07$), commotio cordis ($p = 0.15$), cardiomyopathy ($p = 0.14$), coronary anomaly ($p = 0.8$), and congenital heart disease ($p = 0.054$) did not differ significantly between decease and recovery cases. The proportions of myocarditis (5% and 1%; $p < 0.05$) and aortic dissection (6% and 0; $p < 0.01$) in decease were significantly more than recovered cases.
Conclusions: Although incidence of SCA including both decease and recovery is still around a total of 0.3, approximately two thirds of them have been resuscitated successfully in school from 2008 to 2013. Favorable outcomes in arrhythmic diseases are achieved, however, difficulty of resuscitation in structural disease are enhanced for rescue of SCA in school students.

P2938 - DOES ACTIVATED PARTIAL THROMBOPLASTIN TIME OR HEPARIN LEVEL CORRELATE BEST WITH BLEEDING FOLLOWING SURGERY FOR CONGENITAL HEART DISEASE

Lynn Sleeper[1], Olubunmi Oladunjoye[2], Asha Nair[1], John Kheir[1], Christina Vanderpluym[1], Sitaram Emani[2]
Boston Children's Hospital/Harvard Medical School, Cardiology, Boston-United States[1]; Boston Children's Hospital, Cardiac Surgery, Boston-United States[2]

Background/Hypothesis: Following congenital heart surgery (CHS), patients are at risk for both bleeding and thrombosis. Assays used to monitor anticoagulation with unfractionated heparin (UFH) include the heparin level (anti-Xa) and activated partial thromboplastin time (aPTT). However, disagreement exists regarding which is optimal to guide UFH titration following CHS.
Materials and Methods: We prospectively collected dosing of UFH; aPTT; anti-Xa level; and the presence of major and clinically relevant bleeding in consecutive patients undergoing cardiac surgery and receiving postoperative therapeutic UFH at our institution between August 2015 and July 2016. Major bleeding was defined as that causing emergent surgical intervention or transfusion, intracranial bleeding, or death. Clinically relevant bleeding was defined as that requiring transfusion, imaging or additional testing. All bleeding events were adjudicated. Daily maximum aPTT and anti-Xa values were analyzed using generalized additive logistic regression and logistic piecewise regression with generalized estimating equations for significance testing and estimation of odds ratios.
Results: The analyses of aPTT and anti-Xa were based on 1488 days from 178 patients and 1105 days from 124 patients,

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respectively. Median age at CHS was 2.8 (IQR,0.3-11.4) months and 50% of procedures were RACHS ≥ 4. aPTT and anti-Xa are modestly correlated (R = 0.57). The odds of bleeding are doubled when aPTT exceeds 150 (odds ratio[OR] = 1.71 per 10-sec increase, 95% CI 1.21-2.42; p = 0.003, Figure); however, there is no association as long as aPTT is below 150 (OR = 0.92, 95% CI 0.77-1.10; p = 0.35). Anti-Xa level does not predict bleeding (OR = 1.11 per 0.1 IU/mL increase, 95% CI 0.89-1.29; p = 0.34). *Conclusions:* Following CHS, aPTT exceeding 150 seconds while on UFH is associated with an increased risk of bleeding. Anti-Xa level does not predict bleeding.

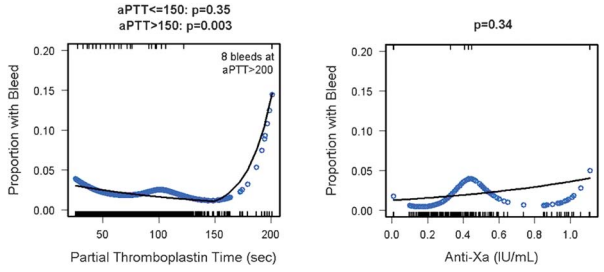

Figure.
Estimated Probability of Bleed when treated post-operatively with therapeutic heparin. Blue curves based on nonparametric model (GAM). Black lines based on parametric logistic regression.

P2946 - CLINICAL PHYSIOLOGIC RESPONSE TO SURGERY AND TO EXTUBATION IN PATIENTS UNDERGOING BIDIRECTIONAL GLENN PALLIATION

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Loma Linda University Children Pediatric Critical Care Medicine Loma Linda-United States[1]; Texas Children Cardiothoracic Surgery Houston-United States[2]; Texas Children Pediatric Critical Care Medicine Houston-United States[3]

Background: Single ventricle anatomy exists in a suboptimal physiological state. The superior cavopulmonary connection results in improved physiology by improving pulmonary blood flow, ventricular volume unloading and improved oxygen saturation (SaO2), and extubation further augmenting pulmonary blood flow. We investigated physiologic changes after the bidirectional glenn (BDG) surgery, as well as the effects of extubation after the BDG.
Methods: We retrospectively reviewed data for patients undergoing BDG over 4 years. Pre and post op ECHOs were reviewed for physiologic response to surgery. Variables averaging 6-12 hours peri-extubation were collected to demonstrate the clinical manifestations of the physiologic response to extubation.
Results: 140 out of 150 patients were intubated post-BDG surgery. Comparing pre-operative to post BDG: greater than mild atrioventricular valve regurgitation (AVVR) decreased from 25.7% to 7.9%, and 13.6% had greater than mild systolic dysfunction with improvement in 2.1%. Comparing pre and post-extubation states, the SaO2 and mean arterial pressure increased (82% to 83%, p = 0.045); (66 to 71 mmHg, p = 0.000) respectively, with an average increase in PaCO2 by 10 mmHg after extubation, and no significant clinical change in PaO2. Post-operative UOP increased, with a decrease in mean pulmonary artery pressure and creatinine post extubation. In 10 patients with an average pre-extubation SaO2 ≤75%, an increase in SaO2 (73% to 76%, p = 0.047), MAP (69 to 74 mmHg, p = 0.011), and post-operative Cr was demonstrated post-extubation.
Conclusion: This large descriptive of patients undergoing the BDG palliation demonstrates clinical trends in certain physiologic variables, with surgery resulting in improvement of AVVR. Extubation improved SaO2, MAP, mean pulmonary artery pressure and demonstrated an average increase in PaCO2 during the post extubation period. We describe these clinical trends in hopes of aiding clinicians gain a better understanding of the physiologic changes after the BDG surgery and clinical response to extubation in this patient population.

P2962 - INFECTIONS IN PEDIATRIC SINGLE VENTRICLE PATIENTS ASSOCIATED WITH INCREASED MORBIDITY AND MORTALITY

Erin Kritz [1], Claire Bocchini[2], Yunfei Wang[3], Paul Checchia[1], Elena Ocampo[4]
Baylor College of Medicine, Pediatric Critical Care Medicine, Houston-United States[1]; Baylor College of Medicine, Pediatric Infectious Disease, Houston-United States[2]; Texas Childrens Hospital, Pediatric Cardiology, Houston-United States[3]; Baylor College of Medicine, Pediatric Cardiology, Houston-United States[4]

Children with single ventricle (SV) physiology represent a vulnerable population with multiple hospitalizations and surgeries. Data is limited on the infectious morbidity and mortality in these patients. Our hypothesis is that SV infants will incur a variety of bacterial and viral infections in the first year of life, which will be associated with worse clinical outcomes. We performed a retrospective chart review of infants with SV physiology born 12/2009 to 4/2015. Of 122 infants, 65% had HLHS; 54% were male; 122 (100%) underwent stage one palliation and 96 (79%) underwent stage two palliation. All cause mortality at one year was 21%. Seventy-six (62%) patients had at least one infection during the first year of life. Thirty patients (25%) had a viral infection, identifying rhinovirus/enterovirus (12), RSV (8), parainfluenza (5), adenovirus (3). The majority (91%) of patients were symptomatic, most often with cough or respiratory compromise. Most viral infections were community-acquired (75%). Infants with viral infections were more likely to have readmission (p = 0.04) and have a heart transplant by one year (p = 0.04). Sixty-two patients (51%) had 149 positive bacterial cultures, identifying 170 organisms from the respiratory tract (64), blood (57), urine (25), other (24). Most common organisms were Enterobacter (29), CoNS (29), Pseudomonas (26), and E. coli (18). The majority (80%) of bacterial infections were nosocomial and symptomatic (88%). Most common symptoms were fever and respiratory compromise. Infants with bacterial infections had longer initial hospital and iCU LOS (both p < 0.01). Infants with bacterial infection prior to Glenn were more likely to have received ECMO (p < 0.01) and to die by one year (p = 0.04). Bacteremia was independently associated with death (p = 0.03). Eight of 26 infants who died had a positive culture in the 48 hours preceding death. Children with single ventricle physiology have frequent infections, which are associated with increased morbidity.

P2966 - A RETROSPECTIVE STUDY OF POST OPERATIVE INTENSIVE CARE MANAGEMENT OF VENTRICULAR SEPTAL DEFECT WITH SEVERE PULMONARTY ARTERIAL HYPERTENSION IN A DEVELOPING COUNTRY

Dr Gopi Nallaiyan
Devadoss Multispeciality Hospital, Paediatric Cardiac Surgery, Madurai-India

Hypothesis: A Retrospective study of Post operative Intensive care management of VSD with severe PAH by a single surgeon form June 2013 TO December 2016 and its results.

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Material and Methods: All cases of surgical closure of VSD with severe PAH was evalvauated restrospectively from June 2013 to December 2016 by a single surgeon. There was 113 cases of Surgical closure of VSD with severe PAH of various age form 4 months to 12 years. All cases were managed with milrinone loading dose during cross clamp release and post operative infusion for 48 hours and then Tab Enalapril 0.2 mg/kg/day and Tab Sildenafil 0.1- 0.2 mg/kg/day in divided doses for 3 months.All patients ventilated with PCO2 of <35mm Hg for 24 – 48 hours with continuous sedation.
Results: 2 mortality out of 113 cases due to PAH crisis and Post operative PA pressures were less than ½ of the systemic in 87 out of 111 cases.
Conclusion: Milrinone is a very good Pulmonary vasodilator for VSD with severe PAH cases in developing nations where there is non availability of Nitric oxide. This is one of the effective way of post operative management in developing countries with financial contriants.

P2970 - PICU OUTCOME OF FULMINANT DILATED CARDIOMYOPATHY AND MYOCARDITIS IN A DEVELOPING COUNTRY

Beyra Rossouw [1], Andrew Argent[2], Brenda Morrow[2], John Lawrenson[2]
Red Cross War Memorial Children's Hospital, Division of Critical Care and Children's Heart Disease, Cape Town-South Africa[1]; Red Cross War Memorial Children's Hospita, Division of Critical Care And Children's Heart Disease, Cape Town-South Africa[2]

Background: Internationally mechanical cardiac support and transplantation have improved long term outcome of children with cardiomyopathy and myocarditis. These treatment modalities are not readily available in the developing world.
Method: A retrospective 7-year single center review of children admitted with fulminant dilated cardiomyopathy (DCMO) or myocarditis to PICU between January 2010 to January 2017 in a setting without available mechanical cardiac support or transplantation.
Results: 112 children with a median age of 27.8 months were included. All presented in Ross stage 4 cardiac failure (77% in cardiogenic shock). Admission median lactate was 6.5 mmol/l. Admission left ventricular ejection fraction was <30% in 79% of patients and 12 developed intra-cardiac clots. Etiology was presumed viral myocarditis in 87% and idiopathic DCMO in 13%. Adenovirus PCR was positive in 28, Parvovirus in 19 with multiple positive viral studies in 32. The median number of PICU admissions per patient was 1.5 (range 1-5) and length of ICU stay was 14.9 days (1-69). 55% required ventilation for median of 8.1 days. 100% required inotropic support for a median of 8.2 days. 82% received Milrinone, 78% Dobutamine and 33% Adrenaline infusions. The median maximum inotrope score was 21.9. Complications during PICU stay included acute kidney injury in 68% of which two patients needed dialysis, liver derangement in 43%, neurological events in 25% and 34% suffered a cardiac arrest episode. 33% had arrhythmias of which 27% needed electrical cardioversion and 57% drug treatment. 63 (66%) children survived to PICU discharge. The overall hospital survival was 47%. Of the PICU survivors the median number of ward readmissions was 3.7 (range 1-19). Total median length of ward stay was 23.2 days (1-138).
Conclusions: In our setting without transplantation availability, DCMO and myocarditis is associated with significant duration of hospital stay, morbidity and mortality.

P2973 - PAEDIATRIC CARDIAC CRITICAL CARE ADMISSION PROFILES AND OUTCOME IN THE DEVELOPING COUNTRY

Beyra Rossouw [1], Andrew Argent[2], Brenda Morrow[2], John Lawrenson[2]
Red Cross War Memorial Children's Hospital, Division of Critical Care and Children's Heart Disease, Cape Town-South Africa[1] Red Cross War Memorial Children's Hospita, Division of Critical Care and Children's Heart Disease, Cape Town-South Africa[2]

Background: In South Africa Paediatric Cardiac Critical Care (PCCC)no uniform PCCC health care model exist. This is the first study to describe a one-year single center South African PCCC patient population.
Methods: A prospective review between January 2015 to December 2015 of all consecutive patients with a cardiac diagnosis admitted to a combined medical and surgical PICU.
Results: 472 PCCC admissions included of which 274 followed elective cardiac surgery, 37 post elective general surgery and 161 emergency admissions. Indications for emergency admission included: shock (28%), respiratory support (22%), cyanosis (18%), decompensated cardiac failure (18%), post cardiac arrest (8%) and life threatening arrhythmias (3%). Comparing elective to emergency admissions the median age was 24 vs 5 months ($p < 0.0001$), length of PICU stay 3 vs 6 days ($p < 0.0001$), length of ventilation 1 vs 2 days ($p = 0.13$), length of inotropes 1 vs 2 days ($p = 0.08$) and median maximum inotrope score 1 vs 9 ($p = 0.46$). A cardiac diagnosis was made for the first time on PICU in 57 patients. 85 emergency theatre procedures were undertaken from PICU. 81 Patients needed PICU readmissions,19 needed redo cardiac surgery. In both groups 4.1% suffered a cardiac arrest during PICU stay ($p < 0.0001$). PICU complications in the elective versus the emergency admission group included failed extubation in 5.7% vs 13% ($p = 0.006$), chest infection on admission in 20.6% vs 45% ($p < 0.0001$), blood stream infection in 3.8% vs 17.4% ($p < 0.0001$), AKI in 17.4% vs 32.9% ($p = 0.001$), liver impairment in 3.5% vs 19.3% ($p < 0.0001$), neurological sequelae in 1.9% vs 14.9% ($p < 0.0001$), PICU readmission in 10.3% vs 30.4% ($p < 0.0001$), PICU mortality 1.6% vs 12.4% ($p < 0.0001$) and standardized mortality 0.25 vs 0.8.
Conclusions: During 2015 34.9% of all PICU admissions were PCCC. Emergency PCCC have a higher morbidity and mortality. This data will be used as benchmarking and towards establishing an outcome based PCCC database.

P3036 - PREVALENCE OF CONGENITAL HEART DISEASE AMONG NEONATES IN A TUNISIAN NEONATAL UNIT OF A TERTIARY CARE HOSPITAL

Jihen Mathlouthi[1], Nabiha Mahdhaoui[1], Houda Ajmi [2], Hedia Ayech[1], Manel Bellaleh[1], Sonia Nouri[1], Hassen Seboui[1]
Farhat Hached Teaching Hospital, Neonatology, Susah-Tunisia[1]; Sahloul Teaching Hospital, Pediatric, Susah-Tunisia[2]

Background: Congenital heart diseases (CHD) have an incidence of 6 to 8 for 1000 live births. Early diagnosis is essential in order to improve prognosis.
Objective: To study the epidemiological and clinical features of CHD in our maternity.
Methods: Retrospective study including CHD diagnosed prenatally or postnatally among the neonates admitted in our department during eight years.
Results: 181 neonates with CHD wère enrolled, 149 of them being inborn among a total of 78,165 live births during the same period (overall incidence of 1.91‰). CHD was diagnosed during first 3 days after birth in 54.7% and before the end of first week in

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64.1% of cases. Diagnosis was suspected prenatally in only 16 patients (8.8%). 159 patients were symptomatic at birth. Symptoms were mainly: presence of an isolated murmur in 39% of cases, cyanosis associated with a murmur in 23.3% of cases, isolated cyanosis in 17.6% of cases and respiratory distress signs in 9.4% of cases. Cardiac ultrasound allowed diagnosis in 179 patients. Non-cyanotic heart diseases with left-right shunt were the most frequent (45.3%) dominated by the interventricular communication (22.65%) and the interatrial communication (11.6%), followed by atrio-ventricular canal (11.05%). Cyanotic heart anomalies due to right to left shunt were present in 28.7% of cases. Tetralogy of Fallot and transposition of the great arteries were the most frequent (8.83% and 7.73% respectively). Obstructive heart diseases observed in 19.9% of the cohort, were dominated by coarctation of the aorta (6.07% CC) and right heart obstructions were dominated by pulmonary stenosis (4.97%). Complex heart malformations represented 6.1% of CHD and were dominated by the single ventricle (4.97%). CHD was isolated in 128 cases. Chromosomal anomalies were found.

P3037 - NEONATES WITH AORTIC COARCTATION AND CARDIOGENIC SHOCK PRESENTATION MANAGEMENT AND OUTCOMES

Jihen Mathlouthi[1], Nabiha Mahdhaoui [1], Houda Ajmi[2], Manel Bellalah[1], Sonia Nouri[1], Hassen Seboui[1]
Farhat Hached Teaching Hospital, Neonatology, Susah-Tunisia[1] Sahloul Teaching Hospital, Neonatology, Susah-Tunisia[2]

Background: Aortic coarctation represents the fifth congenital heart disease in term of frequency. Neonatal forms are generally severe and could be revealed by cardiac failure which is an emergent situation. The aim of this study is to review clinical features, management and short and mid-term outcomes of infants presenting with cardiac failure secondary to aortic coarctation.
Methods: Descriptive study conducted in neonatology department of Farhat Hached universitary hospital (Sousse-Tunisia) during a period of 12 years. Were enrolled all neonates presenting with cardiac failure secondary to aortic coarctation.
Results: Twenty seven neonates were diagnosed with aortic coarctation. Eight of them (29.6%) presented with cardiac failure at the moment of diagnosis. Antenatal diagnosis wasn't performed in any pregnancy. Average age on admission was 10 days, ranging from 7 to 15 days. Continuous perfusion of E1 Prostagnlandines, sodium and fluid restriction associated with vasoactive agents were prescribed to all neonates. Outcome after medical management was good in 5 neonates who underwent surgery after stabilization. Mortality rate was of 50%. Recoarctation was observed in 20% of survivors.
Conclusion: Cardiac failure complicating aortic coarctation is a life threatening situation needing emergent medical and surgical management in order to minimize morbidity and mortality rates. Aortic coarctation screening before decompensation is the best alternative.

INTERVENTIONALCARDIOLOGY

P1022 - REVERSE SZABO TECHNIQUE FOR STENTING A SINGLE MAJOR AORTOPULMONARY COLLATERAL VESSEL IN PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT

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Zaitcev Institute of General and Urgent Surgery, Department of Interventional Cardiology, Kharkiv-Ukraine[1]; University of Texas Health Science Center At Houston, Division of Pediatric Cardiology, Houston-United States[2]; The William Novick Global Cardiac Alliance, Interventional Cardiology Development Coordinator, Memphis-United States[3]; Zaitcev Institute of General And Urgent Surgery, Department of Cardiac Surgery, Kharkiv-Ukraine[4]

Management of pulmonary atresia with ventricular septal defect (PA-VSD) in the neonatal period presents numerous challenges. Endovascular stenting of the ductus arteriosus or of a collateral vessel in ductal-dependent pulmonary circulation as an alternative to the Blalock-Taussig (BT) shunt has become increasingly popular in the last decades. The utilization of the original modification of reverse Szabo (anchor-wire) technique for single collateral vessel stenting in a case of PA-VSD is described.

P1025 - ARTERIAL DUCT STENT IMPLANTATION AMONG DUCT DEPENDENT CONGENITAL HEART DISEASES

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University of the Philippines - Philippine General Hospital, Pediatric Cardiology, San Fernando-Philippines

The objective of the study was designed to determine the outcome of arterial duct stenting in patients with ductal-dependent cyanotic heart disease.
Methods: Between January 2010 and March 2014, 60 cases with duct-dependent pulmonary circulation underwent cardiac catheterization with the intent to do ductal stenting after full assessment by echocardiogram and angiogram. Fifty two cases were included in the study, 22 cases of pulmonary atresia with intact ventricular septum, 12 tricuspid atresia with pulmonary atresia, 9 tetralogy of fallot with pulmonary atresia, 6 double outlet right ventricle with pulmonary atresia, 2 cases of single ventricle physiology with pulmonary atresia and 1 case of tetralogy of fallot with severe infundibulovalvar pulmonary stenosis. All procedures were retrograde through the femoral artery, except four through carotid artery approach.
Results: Twenty five cases were not intubated during the procedure. Twenty six cases underwent balloon atrial septostomy. The mean fluoroscopy time and stay in the hospital from procedure were 15.6 ±11.6 min and 2.7 ±3.3 days. The mean saturation before and after intervention were 62.8%, 85% respectively. Complications pre and post procedure were seen in 21 patients, none of which were via carotid approach. Procedure-related deaths were observed in 7 patients. The causes of death were ductal spasm (n = 6), stent thrombosis (n = 1) and jailed left pulmonary artery (n = 1). One patient expired due to respiratory failure (n = 1), another died a month after discharge due to unrelated cause (n = 1). All 41 cases had stent patency during 4-6 months of follow up. Five among these cases successfully underwent Glenn operation.
Conclusions: Arterial ductal stent implantation is a practicable, effective, safer and less invasive than surgical palliation. Patients with ductal stenting have growth of the pulmonary artery which provides additional time for surgical repair. The carotid artery access is a practical approach among vertical type arterial duct.

P1046 - PERCUTANEOUS BALLOON VALVULOPLASTY FOR SEVERE PULMONARY VALVE STENOSIS IN INFANTS: A 10 YEAR INSTITUTIONAL EXPERIENCE AND LONGTERM OUTCOMES

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Objective: We retrospectively reviewed and analyzed the immediate and long-term outcome and safety of percutaneous

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balloon pulmonary valvuloplasty (PBPV)in neonates and infants with severe or critical valvular pulmonary stenosis.
Methods: One hundred and nine patients aged 2d ~ 3y with critical or severe pulmonary valve stenosis admitted to our hospital from January 2005 to December 2014 underwent balloon valvuloplasty. Right ventricular systolic pressure in all patients was equal to or greater than systemic pressure.
Results: The pulmonary valvuloplasty was successfully performed in 105 of the 109 patients, and the dilatation success rate was 96.3%. Immediately after dilatation, the systemic pressure gradient (PG) from right ventricle to pulmonary artery decreased from 50 ~ 132 mmHg to 4 ~ 96 mmHg. No significant complications in all patients during or post dilation except cardiac tamponade in one. During a 12 months to 9.6 years follow-up (mean 5.01years), data showed that pressure gradient crossing pulmonary valve measured by echocardiography further decreased or remained stable in 103 cases, except one neonate and three infants, whose PG increased and needed a second dilatation (Re-dilatation rate was 3.73%).Mild pulmonary regurgitation was seen in most of patients post-dilatation, except moderate in six and severe in one. All patients were doing well with asymptomatic.
Conclusions: PBPV is safe and effective in attaining both immediate and long term reduction of pulmonary valvular gradients and is currently the preferred therapeutic modality for valvular PS even in small baby patients with severe or critical stenosis.

P1047 - STENTING OF CONGENITAL PULMONARY VEIN STENOSIS A NOVEL HYBRID APPROACH

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Background: Congenital pulmonary vein stenosis is a rare, frustrating and difficult to manage condition with a high mortality rate. New surgical and interventional procedures for treatment suggest an improving prognosis. However, the outcome of patients affected remains unclear and requires often repeated procedures. Our purpose is to present a novel hybrid approach for management of selective pulmonary vein stenosis as alternative to standard percutaneous or surgical repair.
Case: A 6-month-old patient with persistent ductus arteriosus and hypoplastic left pulmonary veins was referred. A failed percutaneous closure of ductus was attempted and surgical closure was then carried out. One month later, he underwent cardiac catheterization for assessment of pulmonary vascular resistances. Two failed attempts for left upper pulmonary vein (LUPV) stenting were performed (left atrium and aortic root perforation resolved with surgical repair). Three days later, he was re-catheterized. A critical diameter (1 mm) of LUPV and left pulmonary wedge pressure of 21 mm Hg were noted. We managed this lesion using a hybrid technique to perforate, dilate and stent the obstructed vessel: the surgical exposure of the Sondergaard's groove was done, and a direct insertion of a bioresorbable stent (3 × 8 mm) using this access controlled by angiography was carried out. The LUPV was then successfully recanalizated. Sternal closure was performed uneventfully.
Results: No significative postoperative complications were detected and he was discharged at home on 11st day. At 18-month follow-up, he is progressing well and echocardiography has shown continuous flow into the stented LUPV and good biventricular function.
Conclusions: This novel hybrid approach should be considered in infants when percutaneous access to the pulmonary veins is challenging or unfeasible. A close partnership of cardiac surgeons with interventional cardiologists is mandatory and it may be the only option for dealing with these lesions in this scenario.

P1050 - IMPLANTATION OF BIOABSORBABLE SCAFFOLDS IN A BABY WITH REPEATED STENOSES OF ALL PULMONARY VEINS

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History: At the age of 3 months a male patient developed severe stenosis of all pulmonary veins. In the cathlab all stenosis were dilated using Paclitaxel covered balloons (IN.PACT Falcon, Invatec, Germany) and conventional balloons (Sapphire, Orbus Neich, Hong Kong). Patient was dismissed home. After three months restenoses occurred. Surgical correction with the sutureless operation was performed and assisted by intraoperative balloon dilation with Paclitaxel covered balloons (Elutax, Aachen resonance, Germany). Again the patient was dismissed home. Three months later he reappeared with hypersystemic right ventricular pressures. Under compassionate care regulations and after having obtained informed consent by the parents, decision was made to implant bioabsorbable Novolimus covered scaffolds (DeSolve, Elixir, USA). Angiography and/or other diagnostic tests: stenosed and stented left lower pulmonary vein.
Procedure: A hybrid approach was chosen to reduce procedure time and radiation. All but the right upper pulmonary veins were found severely obstructed and were consequently stented with 4 ⋆ 18 mm and 3.5 ⋆ 14 mm DeSolve scaffolds under x-ray guidance. After proper placement the delivery material was completely removed and the chest closed. The patient could be extubated after 24 hours and was sent home twelve days later. Medication was diuretics, aspirine and clopidogrel.
Conclusion: Bioabsorbable scaffolds with cytostatic drugs may help to overcome the otherwise often fatal prognosis of intrinsic pulmonary vein stenosis. It rapidly improves the acute life-threatening situation and possibly reduces the chance of restenosis by its continuous cytostatic delivery to the tissue.
Comments: This is a single case with only short term follow-up, so more data and longer follow-up will be needed to allow a conclusion on the possible efficiency of scaffolds in congenital heart diseases. The use of bioresorbable material is still of great interest is all young patients with further growth of their vessels to expect.

P1062 - LONG TERM RESULTS OF BALLON VALVULOPLASTY OF AORTIC STENOSIS

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Background: In years 1987 to 2010, the balloon valvuloplasty was the only method of treatment of congenital aortic stenosis deemed for biventricular circulation in our centre. Long-term results are provided for comparison with emerging surgical methods such as a leaflet shaving and plasty.
Material: Total of 421 consecutive patients were treated at the age of 0 – 23 years (median 109 days, IQR 4 days – 8 years), 141 newborns (33.5%), 105 infants (25.0%), and 175 patients older than 1 year (41.5%). Median follow-up was 10 years (IQR 4 – 16 years).

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Method: Percutaneous valvuloplasty with median balloon-to-annulus ratio of 0.96 (IQR 0.91–1.00). Single-institution regular follow-up. Restenosis defined as Doppler gradient over 70/40 mmHg and severe regurgitation as equal or bigger than the grade 3 of 4. In operated patients, the last values before surgery are used as the latest follow-up.
Results: Before valvuloplasty, after it, and at latest follow-up, the median(IQR) peak Doppler gradient was 85(68-98), 43(35-55), 49.5(35-66) mmHg and the aortic regurgitation ECHO garde was 0(0-0.5), 1(0-2), 2(1-3) (all $P < 0.001$). 57 patients (13.5%) died. 133 patients (31.6%) required surgery. Actuarial probabilities (% ± SEE) at 20 years after the valvuloplasty were as follows: restenosis-freedom 47 ± 4.5, severe-regurgitation-freedom 50 ± 4.6, survival 77 ± 3.8, surgery-free-survival 39 ± 4.4. All the probabilities were significantly lower in newborns than in the other two groups. For the risk factors see Table 1.
Conclusion: Balloon valvuloplasty is a good palliation. Restenosis and severe regurgitation incidences increase over the follow-up period. Risk factors are: small body size, small aortic annulus, LV dysfunction and fibroelastosis, mitral stenosis, and less than 3 aortic cusps.

Table. Cox Proportional Hazard Ratios.

	Restenosis HR	5% CI	95% CI	Severe regurgitation HR	5% CI	95% CI	Death HR	5% CI	95% CI
Fibroelastosis Grade	2.28	1.45	3.56	NS			3.44	1.98	5.96
Mitral Stenosis	2.94	1.65	5.21	NS			3.06	1.61	5.83
Body Surface Area	0.49	0.34	0.71	0.75	0.57	0.98	0.55	0.37	0.82
LV shortening Fraction	0.60	0.48	0.74	0.72	0.72	0.89	0.47	0.36	0.62
Aortic Annulus Z score	0.60	0.46	0.77	NS			0.53	0.39	0.71
< 3 Aortic Cusps	1.94	1.10	3.40	2.35	1.32	4.20	2.10	1.01	4.47

CI - confident interval, HR - hazard ratio, LV - left ventricle.

P1068 - PATTERN OF CONGENITAL HEART DISEASES AND TREATMENT OPTIONS IN A BANGLADESHI CENTER ANALYSIS OF 6914 CASES FROM NONINVASIVE CARDIAC LABORATORY

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Objective: To see the pattern of congenital heart diseases and available treatment facility in a cardiac center.
Background: Echocardiography is the most sensitive tool to diagnose congenital heart diseases efficiently. It can be repeated as many times as required to see the progress, outcome of treatment and complications. Echocardiography laboratory can give a scenario of pattern of congenital heart diseases existing in Bangladesh as well as natural history of the cases and treatment options offered.
Methods: It is a retrospective review of database of echocardiography patient from pediatric noninvasive laboratory of a cardiac hospital in 2012.
Result: Out of 6914 cases 55.73% were male and 44.27% female. Neonates include 8.48%, 36.62% were infant, 34.02% were under 5 years, 17.97% were children and adolescents and 2.96% were adults. Normal Echocardiography finding was found in 17.28% cases of total echocardiography .Ventricular septal defect (VSD) was found in 18.60% cases, Atrial septal defects (ASD) in 15.41% cases, patent ductus arteriosus (PDA) in 8.59% cases, atrioventricular septal defect (AVSD) in 1.97% cases, tetralogy of Fallot (ToF) in 4.79% cases, transposition of great arteries (TGA) in 2.26% cases and Patent Foramen in 265(3.83%) cases. Down syndrome was the commonest chromosomal abnormalities seen in 2.4% cases. Total 599 (8.66%) cases had various types of percutaneous interventions of which PDA device closure was commonest (296 cases). Total 545 (7.88%) cases had various types of surgery of which VSD closure was commonest (107 cases).Four hundred and seventy cases(6.80%) were lost from follow up.
Conclusion: This study proves that VSD is the commonest congenital heart lesions in Bangladeshi children followed by ASD. Frequency of surgery and intervention was almost equal.Some special features of disease pattern were observed in study cases. Some of the cases had spontaneous cure.

P1082 - MID TERM RESULTS OF PERCUTANEOUS VSD CLOSURE WITH ADO II IN PEDIATRIC POPULATION

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Background: Nowadays percutaneous VSD closure is accepted as an alternative surgery but still no ideal device was determined for pediatric population.
Objective: The aim of this study was to share mid term results of Percutaneous VSD closure with ADO II in pediatric population
Methods: VSD closures of 49 patients with ADO-II device were performed in Erciyes University Medical Faculty Children Hospital, Pediatric Cardiology Department.
Results: Mean age of patients: 86.8 ± 52.6months. The youngest patient was 4months old and the oldest patient was 18years old. 19of patients were female, 30were male. Weight of the patients was between 24.3 ± 16 kg (minimum was 5 kg,maximum was76kg). Mean diameter of VSD was 3.7 ± 1,4mm. Mean fluoroscopy and total procedure time were 37 ± 19.3, 74.1 ± 27 minutes respectively. VSD types were muscular in 6patients, rest of the defects were all perimembranous type. No major complications like death, vascular complications, device embolizations was seen. Only complete AV block was seen in one patient month after the procedure and pacemaker was implanted.
Conclusion: Up to our knowledge our study includes the largest series of pediatric patient series that percutaneous VSD closure was done with ADO II. When the complications are taken into consideration in 42 months' follow-up period ADO II device is a good choice in selected cases for VSD closure even in the infants less than 1 year of age.

P1085 - PERCUTANEOUS VSD CLOSURE OF A BABY WEIGHED 3KG

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Objectives: Main aim of our study to share our experience in percutaneous closure of VSD of 3 kg weighed safely with low complication rates.
Background: Surgery is the definitive treatment for ventricular septal defects (VSD) however for muscular defects because of their

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location and coarse trabeculations it is very hard to find and approach these defects.
Case Report: Two months old baby with 3000gr bodyweight was referred to us for tachypnea, tachycardia and growth failure. Pansystolic murmur 2-3/6 in degree was found by physical examination. Apical 4 mm large VSD was detected in the echocardiography, left side of heart was enlarged. 20 mmHg gradient was detected between two ventricles. Her symptoms continued despite anti-congestive treatment. Treatment options were discussed with family. Since VSD is apical located; surgery was found hard and risky. Percutaneous closure was done from arterial route with 5 × 4 ADO II AS (St. Jude Medical, Plymouth, MN) device. We did not face any complications like atrioventricular block during or after the procedure. No vascular complication was seen up till now. In control transthoracic echocardiography no aortic or tricuspid insufficiency was detected. She was discharged on the next day after the procedure.
Conclusion: Up to our knowledge our case is the smallest infant that percutaneous VSD closure was performed successfully in the literature. In this report we emphasized that percutaneous VSD closure is safe even in small infants if appropriate device with appropriate size was used with good technique in experienced hands.

P1124 - CARDIAC CATHETERIZATION IN THE EARLY POST OPERATIVE PERIOD AFTER CONGENITAL CARDIAC SURGERY

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Background: Our aim is to evaluate data of patients who underwent cardiac catheterization in the early post-operative period after congenital cardiac surgery (CHS).
Patients and Methods: We retrospectively evaluated the data of the patients who underwent cardiac catheterization within 30 days after CHS.
Results: 2584 patients were operated in our clinic due to CHD between 2011-2016. Cardiac catheterization was performed to 50 (%1,93) of these patients in early postoperative period. 29 of the patients were males, 21 were females. Median age was 8 years (2 months- 12,5 years) and median body weight was 6 kg (3-35 kg). 56% of the patients had biventricular (n = 28), 44% had single ventricular physiology. Median RACHS-1 score was 3 (1-6). Cardiac catheterization was performed under ECMO support in 16 of the patients. Twenty four (48%) of the patients had only diagnostic catheterization while 26 out of 50 (52%) had invasive procedures. Fifteen patients (30%) were re-operated due to anatomic problems demonstrated during catheterization. Minor complications developed in 7 (14%) patients.
Conclusion: After CHS, during early postoperative period, interventional and/or diagnostic cardiac catheterization can be performed safely to identify and treat problems.

Table 1. The demographic data and the characteristics of the patients.

Patient Characteristics	**N:50**
-Age	8 (2 month-12.5 year) ⋆
-Weight (kg)	6 (3-35)⋆
-Height (cm)	65 (49-150)⋆
-Sex	29 mate/21 female
Postoperative Course	
-Postoperative day of catheterization (days)	12 (1-30) ⋆
-Mechanical circulatory support during catheterization	16 (32%)
-Duration of intensive care unit stay (days)	20 (2-84)
Diagnosis	
-Pulmonary atresia Ventricular septal defect	11
-Right atrial isomerism -Complete AVSD (Unbalanced)	6
-Double outlet right ventricle	5
-Tetralogy of Fallot	4
-Total anomalous pulmonary venous return	3
-Transposition of the great arteries Ventricular septal defect	5
-Shone's complex	3
-Coarctation of the aorta Arcus hypoplasia	3
-Hypoplastic left heart syndrome	3
-Others	7
Indication	
-Prolonged Extracorporeal membrane oxygenation (ECMO)	16
-Prolonged intubation	13
-Low saturation	10
-Glenn dysfunction	2
-Overflow characteristics	2
-Others	7

⋆median (range)

Table 2. Management of the patients.

Intervention	**N:26(52%)**
-Dilation of pulmonary artery	5
-Pulmonary artery stenting	5
-Stent implantation to increase pulmonary blood flow	**4**
-MAPCA embolization	3
-Atrial septostomy	3
-Balloon angioplasty for coarctation and'or balloon dilatation of the aortic valve	2
-Pulmonary vein stenting	1
-Fistula embolization	1
-Mitral balloon angioplasty	1
-Transcatheter VSD closure	1
Reoperation	**N:15(30%)**
-Pulmonary artery repair	**4**
-RV-PA conduit placement repair	3
-Aortic arch reconstruction	2
-Glenn operation	1
-Pulmonary vein repair	1
-Pulmonary artery banding	2
-Glenn takedown	1
-Right atrial maze + fenestration + Pacemaker implantation	1
Change in medical treatment	**N:9 (18%)**

P1125 - ARRHYTHMIAS IN PEDIATRIC CARDIAC CATHETERIZATION LABORATORY

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Introduction: Arrhythmia is a well-known complication of cardiac catheterization in children. Most of them are transient, classified as minor, and don't require treatment. We evaluated arrhythmias emerging during cardiac catheterization in our center.
Patients and Methods: Cardiac catheterization data were collected prospectively under Filemaker® program. Among 2324 patients who underwent cardiac catheterization between January 2012 and November 2016, 143 (6.1%) had arrhythmias during the procedure. Data of these patients were retrospectively reviewed. Arrhythmias during transcatheter ASD/VSD closure, and ST-segment changes were excluded.
Results: In 143 patient, 155 arrhythmia events occurred. Complete AV block (n = 56, 36%) and SVT (n = 36, 23%) were the most common arrhythmias seen. 72(46%) resolved spontaneously in less than 1 minute and 19(12%) resolved just with catheter manipulation. Whereas 35(23%) arrhythmia events needed medical treatment (atropine, adrenalin for AV block and adenosine for SVT) or electrical cardioversion, 26(17%) needed cardiopulmonary resuscitation, one patient with complete AV block needed transient pacemaker, and two others underwent permanent pacemaker implantation one week after catheterization. None of the patients died in the cath-lab and none of them needed extracorporeal life support due to rhythm problems. Diagnostic cardiac catheterization 96/143 (67%) was the most common procedure. Common interventional procedures were pulmonary valve dilation (n = 14), PDA stenting (n = 10). The common cardiac abnormalities were tetralogy of Fallot (n = 27), heterotaxy syndrome (n = 14), double inlet ventricle (n = 14), ccTGA (n = 10). Forty (28%) patients had underwent prior cardiac surgery (palliation or complete repair).
Conclusions: Most of the arrhythmias during cardiac catheterization may resolve spontaneously. But some of them need either catheter manipulation or medication and electrical cardioversion/defibrillation. Life threatening arrhythmias are not as rare as we might think. Pediatric cardiologist should be ready for every kind of arrhythmias during intervention. Judicious catheter manipulation and interfere rapidly to these arrhythmias are crucial to prevent morbidity and mortality.

Table.

	Transient	After catheter manuplation	Medication / Cardioversion	pacemaker	CPR	Total
Complete AV block	32	9	11	3	1	56
SVT	17	6	13			36
2:1 AV blok	8	2	6		1	17
Ventricular tachycardia	3	1			8	12
Atrial fibril as yon/flutter	1	1	4			6
Bradikardi	2		1		2	5
Asistol					10	10
Ventricular fibrilation					2	2
Junctional rhythm	4					4
Elecromecanic disosiation					2	2
Sik PAC/VES	2					2
1. AV block	3					3
Total	72	19	35	3	26	155
%	46	12	23	2	17	100

⋆one patient neededtransent pacemaker, ⋆⋆CPR + defibrillation
CPR: cardiopulmonary

P1144 - TRANSCATHETER CLOSURE OF PATENT DUCTUS ARTERIOSUS 25 YEARS OF CLINICAL EXPERIENCE IN ISTANBUL MEDICAL FACULTY PEDIATRIC CARDIOLOGY DEPARTMENT

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Transcatheter closure of patent ductus arteriosus (PDA) has been suggested to be the standard treatment of PDA.To evaluate the effectiveness of transcatheter closure of PDA this was a retrospective study on patients who underwent transcatheter closure of PDA during the period of 1991–2015. Hospital registry was reviewed and data about patients' characteristics, PDA severity, procedure, and outcomes were retrieved. There were 500 patients, of whom 151 were males, who underwent transcatheter closure of PDA during the study period. Median age was 3.4 years (1 months–18 years), and median body weight was 12 (1.8–65) kg. The diameter of PDA ranged from 1.5 to 10 mm with a median of 2.7 mm. Device could be deployed in all patients, in which most were the coil (52.4%) and Amplatzer ductal occluder (37.8%) and the remainders were umbrella (8.4%), Nit Occlud PDA-R device (1.4%). Complete closure was achieved in most patients (90.3%), minimal residual shunt occured in 8.7% of patients, whereas device migration occurred in (3%) of patients. Coil migration ocurred in 12 patients, 8 patient had bigger than 3 mm PDA and the other 4 patients had smaller than 3 mm PDA. 10 of these migrated coils were retrieved in the same procedure with snare. 2 small coils were not retrieved. All of these patients PDA's were closed in the same procedure with second coil or device. Two patients were not suitable for transcatheter closure and surgery was the choice after the procedure. After closure of PDA with coil, 4 patients had mechanical hemolysis due to residual shunt. All of these patients underwent a second procedure for closure of residual shunt.For closing the residual shunt 1-4 more coils were used. Most patients were discharged from the hospital at 1 day after the procedure.

P1151 - IMPACT OF PULMONARY VALVE REGURGITATION TO EVALUATION OF SEVERITY OF PULMONARY VALVE STENOSIS IN PATIETNTS WITH TETRALOGY OF FALLOT AFTER REPAIR

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Backgrounds: Pulmonary valve stenosis (vPS) and regurgitation (PR) are major postoperative complications in patients with Tetralogy of Fallot. We usually evaluate the degree of vPS by a peak to peak pressure difference between right ventricle and pulmonary artery. However, it is overestimated when PR exists. The purpose of this study is to elucidate the impact of PR to evaluation of severity of vPS.
Materials and Methods: We enrolled 13 patients with vPS without significant PR and 8 vPS patients with PR. First, we examined a relationship between the peak to peak pressure difference and a valve orifice area in patients without PR from their cardiac catheterization data. Secondly, an estimated peak to peak pressure difference, which means the peak to peak pressure difference assuming no PR, was calculated in patients with PR from their valve orifice area using the relationship between the peak to peak pressure difference and the valve orifice area in patients without PR. Moreover, an excess of pressure difference, which means the difference between the measured and estimated pressure difference, was calculated. Lastly, the relationship between the excess of pressure difference and the regurgitant fraction in patients with PR was analyzed.
Results: There was a strong relationship between the valve orifice area and the peak to peak pressure difference in patients without PR ($R^2 = 0.84325$, $P < 0.01$). The excess of pressure difference had a significant correlation with the regurgitant fraction in

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patients with PR ($R^2 = 0.78067$, $p < 0.01$). From this relationship, the PR over 25% of regurgitant fraction could augment the peak to peak pressure difference depending on the regurgitant volume. *Conclusion:* Severity of vPS could be overestimated in post-operative patients of tetralogy of Fallot with PR when it is evaluated by peak to peak pressure gradient, especially if their regurgitation fraction were over 25%.

P1160 - BRAZILIAN SINGLE CENTER EXPERIENCE IN PERCUTANEOUS CLOSURE OF PATENT DUCTUS ARTERIOSUS

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Background: The percutaneous transcatheter occlusion of the patent ductus arteriosus (PDA) had shown great improvement and became a safe method in the last few years. The aim of this study was to describe the experience of a Brazilian Center with percutaneous closure of PDA.
Methods: It was a retrospective single-center study conducted between 2010 to 2016.
Results: From August 2010 to November 2016, a total of 88 patients underwent percutaneous occlusion of the PDA. Sixty patients were female (68%). The mean age was 4.5 years (5 months to 44 years). Based on the Krichenko classification, 59 patients had type A (78.4%), 11 type C (12.6%), 3 type D (3.4%) and 5 type E (5, 7%). In patients with pulmonary hypertension (PH), type C was the most frequent. The most used device was AVP II (52.2%), followed by Coil Flipper (34%), ADO II (5.7%), AVP IV (2.4%). Six patients (6.8%) had associated mild congenital heart disease without hemodynamic repercussion. Immediate complete occlusion was seen in 85 (96,5%) patients. The serial echocardiograms on the follow –up confirmed the initial results. Complications were seen in 5 (5.8%) patients, all in whom were used Coil Flipper, the only device avaliable for PCA closure in our Service until 2005. The embolization of device to left pulmonary artery was the most common complication. Residual shunt after 1 year of the initial closure was seen in 2 patients (2%), in whom were first used a Coil Flipper. In these patients a second closure was perfomed using AVP II, without residual shunt.
Conclusion: In our center, percutaneous treatment of ductus arteriosus was effective and with low risk for the patients. The residual shunt was observed only with Coil Flipper.

P1163 - INTERMEDIATE TERM FOLLOW UP ON THE RESULTS OF TRANSCATHETER CLOSURE OF VSD USING AMPLATZER® DEVICE AND NIT OCCLUD® LEˆ COIL

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Introduction: Transcatheter ventricular septal defect (VSD) is alternative treatment of VSD closure especially in perimembranous type (Pm VSD) and doubly committed subarterial (DCSA) VSD.
Material and Methods: The aim of this retrospective study is to review intermediate term (up to 5-years) results of transcather closure of VSD using Amplatzer® device (device) or Nit Occlud® Leˆ VSD Coil (coil).
Results: 240 out of 247 patients (97.1%) underwent transcatheter closure successfully (187-device and 53-coil). The median age and weight were 12 years (1-67) and 40.3 kg (10-97), respectively. The median VSD size in device group was larger than coil group. [7 mm (3-18) VS 5 mm (2.5-9.3), $p < 0.001$]. Complete closure was comparable among both groups. Twenty-six patients (49.1%) in coil group were DCSA VSD while only 42.2% in device group. Sixty-nine (26%) of all patients had DCSA VSD (43-device and 26-coil). Although pre-closure aortic regurgitation (AR) was comparable on both groups (22.9% VS 32%, $p = 0.448$). AR were significant higher in coil group ($p < 0.001$) than in device group. Comparing VSD type, AR post procedure was higher in DCSA VSD than Pm VSD ($p = 0.001$). At the end of 5-year FU, 98.6% of DCSA VSD had less than mild AR.
Conclusions: We found comparable closure rate by using either device or coil for transcatheter closure of VSD in both Pm VSD and DCSA. The progression of AR was higher in DCSA VSD patients but majority of them had less than mild AR at end of 5-year follow up.

P1169 - BALLOON ANGIOPLASTY FOR COARCTATION OF AORTA IN INFANTS

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Background: Few studies have compared the outcomes of balloon dilatation for post-surgical and native coarctation in infancy .This study evaluated procedural outcome,reintervention and late clinical outcomes of balloon angioplasty in infants with post-surgical recoarctation and native coarctation .
Materials and Methods: Retrospective case sheet analysis of 35 consecutive infants (Group A: post surgical re-coarctation; Group B: Infants above 3 months with native coarctation), who underwent a total of 45 procedures of Balloon angioplasty during 2011-2015. Results are shown in Table 1(Figures with ± denote mean ± standard deviation.
There were no major complications. One baby(group B) did not respond and underwent surgery. 7/36 babies had femoral artery occlusion requiring heparin infusion. Clinical/echo follow up was available for 34 patients, additional imaging in 21 patients.

Table.

	Group A (n = 25)	Group B (n = 10)	p value
Age at surgery, days	11.5 ± 9.19	-	-
Time to balloon, months	5.50 ± 0.71	-	-
Age at first cath, months	5.92 ±2.55	5.90 ± 2.69	0.98
Weight at first cath, kg	6.04 ± 2.18	5.94 ± 1.63	0.90
Arch hypoplasia, number	10/25	1/10	-
Balloon/aorta ratio	0.99 ±0.18	0.91 ± 0.19	0.24
% increase in diameter	63.24 ± 35.46	61.18 ± 39.51	0.88
% drop in gradient	54.00 ± 20.75	49.10 ± 32.17	0.60
Recath, number	12/25	4/10	0.72
Follow up , months	34.00 ± 33.94	24.00 ± 25.46	-
BP > 95th percentile	7/24	1/10	0.39
Upper-lower limb BP gradient > 15 mmHg	4/24	2/10	1.00

Figures with ± denote mean ± standard deviation

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Localized aneurysm was noted in one. Intervention for recurrent CoA (n = 10)showed increase in diameter of 61.33 ± 35.68% and drop in gradient t of 57.22 ± 23.38%. Nine babies underwent planned surgery/intervention for associated lesions. 27/35(77%) children were free of hypertension and/or significant BP gradients between upper and lower limbs .
Conclusion: Infants with post-surgical recoarctation and selected cases of native coarctation respond to balloon angioplasty with good procedural outcomes and similar freedom from hypertension in childhood.

P1170 - ATRIAL SEPTAL DEFECT DEVICE CLOSURE IN CHILDREN WEIGHING LESS THAN TEN KILOGRAMS

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Background: Trans catheter device closure is the standard of care in children with ostium secundum ASD.The procedure is generally deferred till the child is about 15 kg. Selected children do need to undergo ASD device closure at an younger age. This study evaluated the safety and efficacy of the procedure in these children and the impact of the procedure on their growth.
Material and Method: Retrospective case sheet analysis of 14 consecutive children weighing <10 kg who underwent device closure of secundum ASD during the period January 2011 through December 2015,for heart failure, failure to thrive or recurrent respiratory infections.
Results: Results are shown in Table 1.
There were no major complications. One patient had transient complete heart block while another had mild SVC obstruction (Doppler mean gradient = 3 mmHg). At a mean follow-up of 22.45 months (±18.18) the weight for height Z-score of the group improved marginally from -2.96(±1.98) to -2.53(±1.51). The proportion of children having a Z-score more than -2 dropped from 11/14 (78.5%) to 6/11(54.5%). Failure to gain weight was associated with major co-morbid conditions including chromosomal anomalies (n = 4) and multi-organ anomalies (n = 1).
Conclusion: When indicated ASD device closure can be performed safely in children weighing less than 10Kg with excellent long term outcome. Weight gain following the procedure is adversely affected by associated chromosomal syndromes or multi-organ anomalies.

Table.

PARAMETER	MEAN(± SD)
Age (months)	20.92 (±13.73)
Weight (kg)	7.83 (±1.61)
Wt./Height Z-score	-2.96 (±1.98)
Fluoroscopy time (mts)	9.66 (±6.56)
Radiation (mGy)	40.50 (±32.72)
Device size (mm)	13.14 (±3)
ASD size (mm)	11.98 (±2.83)
ASD size/Weight ratio	1.59 (±0.53)

P1202 - TRANSCATHETER INTERVENTION FOR COR TRIATRIATUM IN 41 YEAR OLD MALE

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Forty one year old gentleman was referred to cardiology when he started to experience symptoms of breathlessness and effort intolerance. Transthoracic echocardiogram revealed clear membrane in left atrium with high velocity flow suggestive of obstructive Cortriatriatum. Estimated pulmonary arterial pressures were systemic. Cardiac MRI confirmed Cortriatriatum with small perforation and ascending vein communicating between left superior pulmonary vein and left innominate vein resulting in left to right shunt quantified at 3:1. Cardiac multidisciplinary meeting decided primary surgical repair would carry an increased risk because of longstanding pulmonary hypertension and balloon dilation of the Cortriatriatum membrane as initial palliative procedure to try and reduce his pulmonary pressures would be an appropriate step. In Catheter Lab, pulmonary artery pressures were near systemic (77 systolic compared with femoral arterial systolic 85 mm Hg). The Cortriatriatum membrane was ballooned with 25 × 45mm Crystal balloon, inflated to 4 atmospheres pressure on three occasions. Each balloon inflation abolished the waist but this returned on deflation. Transoesophageal echo showed the Cor orifice was enlarged by approximately 50%, however it was unclear if this was adequate as pulmonary artery pressures remained high due to persistent significant left to right shunt through the ascending vein. We therefore abandoned the procedure and the patient went for surgery. Intraoperatively, the Cor membrane was found to be torn, this was resected and ascending vein ligated. Recent follow-up after surgical repair, patient is well with no symptoms and marked improvement in his estimated right heart pressures seen on transthoracic echocardiogram.
Conclusion: The transcatheter intervention was partially successful, however the result of balloon dilation was difficult to quantify as pulmonary artery pressures remained high due to significant left to right shunt through ascending vein. We believe balloon dilation could be considered as a palliative or corrective option when primary surgical repair is high risk.

P1213 - PULMONARY VASCULATURE REMODELING IN A HYPOPLASTIC LEFT HEART PATIENT

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Background: Patients with hypoplastic left heart syndrome (HLHS) and restrictive atrial septal defect (ASD) may benefit from fetal septostomy.
Objective: We assessed the extent of pulmonary vascular lesions in a newborn diagnosed with HLHS and restrictive ASD, from birth to the 26th day of life.
Methods: Patient was submitted to intrauterine balloon septostomy at the 28th gestational week. At birth, obstruction to left atrial egress was observed on echocardiogram. Immediate hybrid procedure was carried out, with stents implanted in the atrial septum and the patent ductus arteriosus. A lung biopsy was harvested on day 1 of life. The patient evolved with progressive signs of

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pulmonary congestion due to persistent restriction of the ASD. On the 19th day of life, atrial stent ballooning did not relieve pulmonary venous congestion. Atrial septal resection under cardiopulmonary bypass support was performed on the 26th day of life. A new lung biopsy was collected. Patient did not survive the operation.
Results: After conventional histological processing, the lung biopsies were evaluated by qualitative analysis and morphometry, with calculation of the percentage of arterial medial thickness. The first biopsy (Day 1) showed diffuse and moderate hypertrophy of the arterial medial layer (Rabinovitch grade B), lymphatic dilation and venous wall thickening. The percentage of arterial medial thickness varied from 0.71 to 1.66 times the normal reference for age (mean Z value = + 0.65). Histologic changes of the second biopsy (Day 26) were more severe. Congestive pulmonary findings persisted. Severe occlusive intimal lesions were noticed in both pre and intra-acinar arteries. The medial thickness was increased up to 3.55 times the normal reference value (mean Z value = + 1.74).
Conclusion: Pulmonary vascular lesions progressed rapidly after birth due to unsuccessful repeated attempts of ASD enlargement. Probably, early relief of the obstruction would be more efficient to improve survival of these patients.

P1233 - CHARACTERISTICS AND OUTCOMES OF CHILDREN WITH TAKAYASU'S ARTERITIS AT A SOUTHERN AFRICAN TERTIARY CARE CENTER

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Introduction: Takayasu's Arteritis (TA) is a rare granulomatous, fibrosing arteritis of the aorta and its branches resulting in stenosis, occlusion, dilatation or aneurysm formation. Hypertension (HT) and reduced or absent pulses are common manifestations. A dilated cardiomyopathy (DCMO) is often contributed to HT, mitral and or aortic valve disease but myocardial involvement itself may be the cause. The aetiology is unknown, but tuberculosis (TB) and autoimmune aetiologies have been suggested. Management includes anti-inflammatory and chemotherapeutic agents, and revascularization.
Objective: To assess the characteristics and outcomes of patients with TA at a large Southern African Tertiary care centre.
Methodology: Retrospective analysis of all children diagnosed with TA over a 20 year period.
Results: A total of 55 children were documented with an average age of 9.7 years (SD, 3.4), F:M ratio 3:1, involvement of the abdominal aorta 34/55 (62%), renal artery stenosis 35/55 (64%), positive TB skin test recorded in 40/55 (73%) and a DCMO in 37/55 (67%) with an average shortening fraction (SF) of 20% (SD, 11%). The majority manifested with HT 39/55 (71%), heart failure in 24/55 (44%), and seizures in 15/55 (27%). All patients were treated with steroids, chemotherapeutic agents (cyclophosphamide and methotrexate), and TB treatment. Twenty three patients underwent 30 percutaneous revascularization procedures to treat uncontrolled HT: 8 stents (aorta 5, renal arteries 3), 22 balloon angioplasties (aorta 6, renal arteries 15, branch pulmonary artery 1). The systolic blood pressure decreased from 153 ± 23 to 133 ± 24 mmHg ($p = 0.005$). All patients required continued antihypertensive treatment after intervention. The SF improved in both the intervention and non-intervention groups.
Conclusion: TA at our institution is commonly associated with HT, heart failure and seizures. Percutaneous intervention, although not curative, achieves better control of HT. Reduced left ventricular function improved in all patients receiving medical and/or percutaneous interventional therapy.

P1237 - 3D ROTATIONAL ANGIOGRAPHY WITH 3D ROADMAP GUIDANCE IN CHD INTERVENTIONS

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Background: Nowadays, interventional cardiac catheterization has become an important component of the management for congenital heart diseases. Important advances in imaging modalities such as 3D rotational angiography (RA) with 3D roadmap guidance (RG) allows for both advanced diagnostic and interventional cardiac catheterization procedures. Recently, in our institute 3D RA with 3D RG was available first in Asia. This new technology is tremendously helpful in cardiac interventional procedures, that is, easier procedure, reducing angiography, radiation exposure and contrast amount. We would like to share our initial experiences with the audience.
Method: Between November 2013 and January 2016, 3D RA with 3D RG was used in 27 CHD interventions.
Results: 3D RA with 3D RG was used for pulmonary arterial interventions in 23 patients: congenital multiple peripheral pulmonary stenosis (13), postoperative pulmonary stenosis (8), and Fontan (2), stent for coarctation of aorta in 2 patients, and embolization of pulmonary arteriovenous fistula in 2 patients. Mean age was 13 ± 5 years, and mean body weight was 41 ± 20 kg. The number of target vessel was 2.3 ± 1.7. Mean contrast amount for 3D RA was 1.2 ± 0.5 ml/kg. Fluoroscopic time and procedure time were 32 ± 17 and 128 ± 35 min., respectively.
Conclusions: 3D RA with 3D RG is a promising modality for CHD diagnosis and treatment. 3D roadmap overlaid onto live fluoroscopy is tremendously beneficial for some CHD interventional procedures. It reduces contrast dose and procedural time.

P1256 - DRUG COATED BALLOON ANGIOPLASTY FOR PULMONARY VEIN STENOSIS IN PATIENTS WITH RECURRENT PULMONARY VEIN STENOSIS AFTER SURGERY INITIAL EXPERIENCE WITH A NEW TREATMENT TECHNIQUE

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Background: Pulmonary vein stenosis (PVS) is a serious complication following TAPVR repair. The main therapies for PVS include surgical repair, balloon angioplasty (BAP) and stenting, but these treatments are complicated by a high rate of restenosis. We describe the use of a paclitaxel drug-coated balloon(DCB) for the treatment of recurrent PVS.
Materials and Methods: We used 4mm DCB (SeQuent Please, B. Braun, Hessen, Germany) to treat PVS for 3 patients with recurrent PVS after TAPVR repair, surgical PVS release, and conventional BAP. For 2 patients, we used conventional 5mm high-pressure PTA balloon in advance, then introduced DCB, and for 1 patient, used DCB from the beginning. We performed follow-up angiography 2 months later in 2 patients, 6 months later in 1 patient.
Results: Follow-up angiography showed effectiveness of DCB for the reduction of restenosis compared to the conventional BAP underwent previously for the same lesions in all 3 patients. All patients showed no complication and no adverse effect in 6 months follow-up period.

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Conclusions: Combination therapy using high-pressure PTA balloon and DCB for recurrent PVS after TAPVR repair is feasible and safe. This novel strategy may postpone the timing to perform next intervention and provide good long-term results for patients suffering from a serious and life-threatening complication after surgery.

P1296 - IS BALLOON AORTIC VALVOTOMY STILL THE PREFERRED INTERVENTION IN NEONATES AND INFANTS WITH CONGENITAL AORTIC STENOSIS SINGLE CENTER EXPERIENCE AND LITERATURE REVIEW

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Neonates and infants with congenital aortic stenosis are hemodynamically fragile. While most institutions, prefer balloon aortic valvotomy (BAV) for treatment, few centers have demonstrated excellent outcomes with surgical valvuloplasty (SAV). We studied our immediate and mid-term results for BAV in these patients.
Methods: Retrospective records review of neonates and infants who underwent BAV at our center from 2010 to 2015: baseline characteristics, procedural details and follow-up variables analyzed.
Results: 26 infants (6 females, 15 neonates) studied. Median age 1 month [2 days - 10 months]. Mean weight 3.92+ /- 1.36 kg. 58% patients had significant LV dysfunction (34.6% severe) at presentation. Mean aortic annulus during BAV 7.1 mm + /- 1.2. Mean balloon width to aortic annulus ratio 1.04 + /- 0.25. Mean AS gradient 60.9 + /- 14.9 mm Hg. Mean post valvotomy gradient 19.2 + /- 13.6mm Hg. Mean reduction in AS gradient 43.8 + /- 15.9 mm Hg.
Immediate post valvotomy, 5 (19%) moderate AR, 10 (38%) mild AR, 11 trivial/no AR; 3 (11.5%) had mild dysfunction, 2 (7.7%) severe dysfunction. Median hospital stay 2 days [1-22 days]. Median follow-up duration 54 months [6–76 months]. 6 patients (23%) required repeat BAV; median interval of 6 months [0.8-24 months]. Predictors of reintervention: insufficient decrease in gradient and post-valvotomy gradient > 30 mm Hg.
No immediate mortality. 1 child expired during reintervention (3.8%). None of our patients required surgical reintervention till date. Mortality and reintervention was significantly lower than reported in literature. Repeat balloon valvotomy required in only 23%.
Conclusion: Balloon aortic valvotomy can achieve good immediate and mid-term results in neonates and infants with very low procedural and early mortality and morbidity. Present results support BAV as the preferred intervention in these patients.

P1398 - EARLY AND LATE PRESENTATION OF CORONARY ARTERY FISTULA AN EXPECTED NATURAL PROGRESSION

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Background: Coronary artery fistulas (CAF) are rare. They arise proximally or distally and most often drain to the right heart. They do not usually cause symptoms or complications in childhood, especially when small. However, symptoms and complications occur frequently in adults and include congestive heart failure, myocardial steal, thrombosis, myocardial infarction, stroke, arrhythmias, and sudden death.
Hypothesis: CAF if untreated in childhood, undergo a natural progression from the chronic shunts that create more complications in adults.
Materials and Methods: Two patients are compared with the same CAF. A 10 yo girl was diagnosed as a newborn, was asymptomatic for many years, and recently developed exertional chest pain. Echocardiogram showed a dilated left main coronary artery (LMCA) and large proximal CAF draining to the RA. Also, a 43 yo lady presented with a yearlong history of recurrent extremity numbness, palpitations, and dyspnea on exertion. Echocardiogram showed an enlarged LMCA with a large, tortuous CAF. Coronary CT angiography (CTA) confirmed a large proximal CAF draining to the RA.
Results: In both instances, the CAF arose from a large LMCA proximal to the bifurcation of anterior descending and posterior circumflex branches, turning sharply posteriorly and rightward to course tortuously behind the aorta and enter the right atrium superiorly. The CAF in the adult was significantly larger, as expected. CTA provided important anatomic detail to help plan a treatment strategy. Transcatheter device closure was successful in both, but challenges occurred due to tighter catheter turns in the child and due to the very generous CAF diameter in the adult, who had a complicated post-procedure course.
Conclusions: CAF closure is indicated when symptoms exist. Delay in treatment until symptom onset can create complications and a more challenging substrate for successful intervention. Closure should be considered at younger ages to minimize later complications.

P1400 - USE AND SAFETY OF CARDIAC CATHETERIZATION PERFORMED IN POST NEONATAL HEART SURGERY WITH POOR HEMODYNAMICS EVOLUTION. STUDY IN A CENTER OF MEDIUM COMPLEXITY

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Background: When a persistent and severe hemodynamic decompensation occurs in the immediate postoperative of infants with systemic pulmonary shunt surgeries, patent ductus arteriosus and coarctation of the aorta, it raises the use of diagnostic methods that exceed the clinical analysis and echocardiogram. In these cases, the cardiac catheterization study is the standard for diagnosis and occasionally, also therapeutic.
Material and Method: We analyzed the 498 cardiac catheterizations performed in the last five years. Twenty-four procedures were in severe and persistent hemodynamic compromise after neonatal postoperative in this group of patients.
Results: In the systemic pulmonary shunt surgeries were found: A) Severe stenosis of the subclavian artery: five cases and we dilated the subclavian artery in 2 cases with balloon alone and in 2 with stents. B) Stenosis of the pulmonary arteries: 3 cases. C) Shunt thrombosis/Thrombolitic therapy: 2 cases. D) Large aorto-pulmonary collateral: 1 case. E) One shunt anastomosis from the subclavian to an unobtrusive total anomalous pulmonary venous collector. F) Giant pseudoaneurysm: 1 case. In coarctation of the aorta: A) Severe stenosis in surgical repair: 4 cases. Dilation the recoartation in 3 cases and dilation of ductus in 1. B) Hypoplasia of the aortic arch: 2 cases. In closing patent ductus arteriosus: A) Pulmonary sequestration: 1 case with posterior embolization B) Hepatic arteriovenous malformation 1 case C) Thrombosis in superior vena cava with posterior percutaneous transluminal angioplasty: 1 case. In two patients the anatomy of the surgery was the correct one. No patient died during the diagnostic or the therapeutic procedures.

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Conclusions: Diagnostic and therapeutic cardiac catheterizations are useful and safe in this group of patients.

P1402 - OUTCOMES OF RADIOFREQUENCY PERFORATION FOR PULMONARY ATRESIA AND INTACT SEPTUM VENTRICULAR SEPTUM AND CRITICAL PULMONARY VALVE STENOSIS WITH CORONARY FISTULAS. A SINGLE CENTER 20 YEARS EXPERIENCE

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Background and Aims: Newborns with Pulmonary Atresia with intact ventricular septum (PAIVS) and Critical Pulmonary Stenosis (CPS) are cyanotic at birth and require prostaglandins (PGE) for maintenance of pulmonary blood flow. These patients' spectrum is wide, ranging from those with relatively good-sized RV chambers to those with significant RV hypoplasia or RV-dependent coronary circulation. The presence of coronary fistulas has been classically a contradindication for opening the pulmonary valve.
The main objective of this study is to review our 20-year experience in managing PAIVS and CPS, to analyze early and long-term results and the main decision features toward a biventricular or a univentricular pathway with the strategy of opening the valve although the presence of coronary fistulas.
Methods: This is a descriptive retrospective study where we identified all patients diagnosed with PAIVS or CPS admitted to the neonatal intensive care unit (NICU) between January 1996 and October 2016. For each patient, available medical records, echocardiograms, angiograms, and operative reports were reviewed. Morphologic RV features, coronay fistulas, surgical management, percutaneous intervention and long-term outcome data were collected and analysed.
Results: 52 patients were admitted. Major features between both groups can be seen in Table 1. A total of 9 (17.3%) patients died during the follow-up period. 8 APSI and 1 CPS. None of the patients with coronary fistulas died. The 43 survivors were followed up for 107.81 months (±53.56). New York Heart Association functional classification at the most recent examination was for the overall group 1.2, being 1.2 for the ones with BV, 2 for the UV and 1.3 for the 1.5 V. Regarding the CHD SPS survivors had a functional class of 1.1 and the PAIVS a 1.6.
Conclusions: Strategy of aggressive and precocious early opening of the pulmonary valve has a good global survivence correlated with

Table 1. Patients' clinical characteristics data.

Variable	CPA (n = 23)	PAIVS (n = 29)	p value
Female sex	12 (52.2%)	15 (51.7%)	0.624
GA weeks ± SD	37.7 ± 3.8	38.7 ± 2.0	0.794
Prematures (<37 GA weeks)	4 (17.4%)	4 (14.2%)	0.316
BW (Kg) ± SD	2.85 ± 0.80	3.11 ± 0.59	0.337
SGA	3 (13.0%)	5 (18.5%)	0.885
Prenatal Diagnosis	15 (65.2%)	24 (86.2%)	0.042
Extracardiac malformations	1 (4.3%)	6 (20.6%)	**0.037**
RV partite status			
Tripartite	19 (82.6%)	22 (78.6%)	0.623
Bipartite	4 (17.4%)	5 (17.2%)	0.756
Unipartite	0	2 (7.1%)	**0.021**
TV Z score	-1.73 ± 0.62	-2.17 ± 0.85	0.08
RVOT Z score	-2.65 ± 1.18	-3.64 ± 1.63	0.17
Coronary fistulas	2 (9.1)	6 (21.4)	0.23

CPS: Critical Pulmonary Stenosis. PAIVS: Pulmonary atresia with intact ventricular septum. GA: gestational age. SD: Standard Deviation. BW: Birth weight. Kg: kilograms. SGA: small for gestational age (BW < p10). TV: tricuspid valve. RVOT: right ventricular outflow tract. Significant values are in bold numbers.

P1409 - PROCEDURAL OUTCOME AND SHORT TERM FOLLOW UP OF PATIENTS UNDERGOING ENDOVASCULAR STENTING FOR COARCTATION OF AORTA A SINGLE CENTRE EXPERIENCE

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Background: Endovascular stenting is the preferred option in managing coarctation of aorta (COA) in older children and adults. Covered stents are used in selected or high risk catergory of patients.
Materials and Methods: Patients with severe coarctation of aorta who underwent endovascular stenting during the period May 2013 to November 2016 were retrospectively analysed. CT aortogram was used for preprocedural imaging.Procedural outcome complications and short term follow up were noted.
Results: 26 patients (eight females) aged 1-54 years (median 29), weighing 7.8 -86.4 (median 55.1) kg, underwent stenting of COA. All except one had postsubclavian coarctation. 70% of patients had hypertension and were on treatment. Mean gradient at catheterisation was 69.2 ± 29.4 mm hg and mean gradient postprocedure was 3.5 ± 4 mmhg. A total of 27 stents were deployed, Covered CP (17), Cook Formula (1), Advanta V12 Atrium (2), Intrastent Mega (3), Palmaz (2), Andrastent. (1), Bare CP (1). Covered stents were used in 59.1%. The mean stent length and balloon diameter were 34.73 ± 11.56 mm and 16.3 ± 2.42 mm respectively. Pre dilatation was done in two patients including one with near interruption. Post dilatation was needed in 35% of patients. Procedural complications included dissection in one patient needing a second covered stent and right femoral artery occlusion in another. Retroperitoneal hemorrhage in one resulted in mortality. Follow-up ranged from 1 month to 3.5 years. 36.4% required continuation of antihypertensive therapy even after stenting. One patient with presubclavian coarctation required redilatation 6 months after stenting.
Conclusions: Stent implanation is a safe and effective alternative to surgical repair in COA. It provides immediate and near complete relief of obstruction which is sustained on short term follow up. Long term follow up is required to look for restenosis, aneurysm formation and persistent systemic hypertension.

P1410 - STENTING OF RIGHT VENTRICULAR OUTFLOW TRACT CONDUITS AND PULMONARY ARTERIES – PROCEDURAL OUTCOME AND MEDIUM TERM FOLLOW UP

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Background: Stent angioplasty is an accepted treatment for narrowed pulmonary arteries(PA). It also postpones surgical conduit replacements and offers landing zones for percutaneous pulmonary valves.

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Materials and Methods: Retrospective analysis of patients who underwent PA and RVOT stenting over a period ranging from June 2008 to June 2016. Patients were divided into Group A with elevated right ventricular(RV) pressures due to narrowed conduits, main PA or bilateral branch PA and Group B with unilateral PA stenosis. Their etiology, outcome complications and short term follow up were analyzed.
Results: Among 80 patients(39 males) between 0.25-37 years (mean 10), weighing 4.5-82(mean 27) kg divided into 46 in Group A and 34 in Group B, 73 patients had prior surgery 3 days to 17 years earlier.Conduit stenting in 20 patients were successful in all, uniformly postponed their replacements and offered landing zones for percutaneous valves in six. One among the four MPA stenting did not give adequate result and required surgery. Bilateral PA stenting procedure was successful in all 22 patients, often needed simultaneous deployment. On a five-year follow-up, five needed reinterventions. Unilateral PA stenting was successful in 32 out of 34 patients, one died of vessel rupture and other needed surgery for embolization. Stent migration in two and fractures in two needed additional stent placements. There were eight complications (10%)in total; young age and fresh suture lines were risk factors.
Conclusions: Stenting postponed surgical replacement in stenosed conduits and served as landing zones for percutaneous pulmonary valves. Bilateral PA stenting was uniformly effective in reducing RV pressures, and often needed simultaneous deployment. As reinterventions might be needed with somatic growth, the stents should be post-dilatable. In discrete LPA origin stenosis, short stents and stents with poor radial strength should be avoided to prevent stent migration and fractures.

P1416 - COMPARISON OF CARDIAC INDEX MEASUREMENTS BY CATHETRIZATION AND PHASE CONTRAST MRI IN CHILDREN WITH PULMONARY HYPERTENSION

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Background: Hemodynamic evaluation via catheterization is essential for diagnostic and therapeutic investigation of children with pulmonary hypertension (PH). Measurement of pulmonary flow is required to calculate pulmonary vascular resistance index (PVRi), an important component of investigation and a predictor of outcome. Calculation of cardiac (CI) index using the Fick principle in most catheter laboratories relies on an estimate of oxygen consumption which may limit its reliability. In this study, we compared CI assessed by the Fick principle and thermodilution (TD) with gold-standard flow waveform evaluation technique – phase-contrast MRI (PC-MRI).
Materials and Methods: Twenty-four patients with pre-diagnosed idiopathic PH (n = 19) or PH associated with congenital heart disease (n = 5) (age: 14.2 ± 4.6 (range: [4, 20]), M:F = 10:14) underwent same-day right-heart catheterization and PC-MRI for hemodynamic evaluation. CI computed from the catheterization (Fick and TD) were compared with PC-MRI (indexed integrated flow waveform multiplied by heart rate) using linear regression models (beta ± standard error) and Bland-Altman analysis.
Results: CI measured by the Fick principle and PC-MRI failed to show significant correlation (0.06 ± 0.24, p = 0.8030). Furthermore, no significant correlation existed between TD and PC-MRI (0.78 ± 0.44, p = 0.1080). Respective Bland-Altman analysis revealed a bias in CI of -2.8 and -2.4 L/min/m2 between Fick vs. PC-MRI and TD vs. PC-MRI, respectively. The limits of agreement for the Fick method vs. PC-MRI were -2.6 to 2.4 L/min/m2 and -2.1 to 2.1 L/min/m2 for TD vs. PC-MRI.
Conclusions: CI calculations differ significantly between catheterization and PC-MRI. Stronger agreement existed between TD vs. PC-MRI than between Fick vs. PC-MRI. The Fick calculation and thermodilution techniques have well recognized limitations and errors; however PC-MRI assessments are not without their limitations. Future studies are required to evaluate the relative prognostic value of PVRi and CI calculated using different modalities.

P1428 - INITIAL EXPERIENCE OF TRANSCATHETER CLOSURE OF SUBARTERIAL VENTRICULAR SEPTAL DEFECT WITH THE AMPLATZER DUCT OCCLUDER

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Background: Traditional treatment of subarterial ventricular septal defect is open heart surgery. This study aimed to evaluate the feasibility, safety and outcome of transcatheter closure with the Amplatzer duct occluder.
Methods: Between March 2012 and June 2015, a total of 16 patients with subarterial ventricular septal defect who underwent transcatheter closure with the Amplatzer duct occluder were enrolled retrospectively. There were 8 males and 8 females. The age ranged from 3.0 to 65.6 years with the median of 35.6 years. The body weight ranged from 14 to 92 kg with the median of 60 kg. All patients had a prolapse of right coronary cusp without subaortic rim. Mild aortic regurgitation was noted in 11 (69%) patients.
Results: Left ventriculogram showed ventricular septal defect size ranged from 1.3 to 9.3 mm with the median of 3.5 mm. Device was successfully implanted in 88% (14/16) patients. One patient had mild skin allergy to contrast medium. No other complication was noted. Complete closure rate was 64%, 86%, 86% and 86% at 1-day, 1-month, 6-month and 12-month follow-up respectively. Two patients developed new or worsening aortic regurgitation during follow-up.
Conclusions: Transcatheter closure of subarterial ventricular septal defect with Amplatzer duct occluder is technically feasible and safe in patients older than 7 years old. However, development or worsening of aortic regurgitation needs long-term follow-up.

P1449 - PERCUTANEOUS VSD CLOSURE UNDER 1 YEAR OF AGE

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Background: Untreated large Ventricular septal Defects (VSD) are important reason of congestive heart failure in early infancy. This population usually fails togrow and surgical closure is challenging because of congestion in their lungs prone to respiratory infection, their bad nutritional status.
Objective: The aim of this study was toshare our experience in VSD closure of children under 1 yearof age percutenously.
Methods: We have performed VSD closure of 7patients under 1year of age between the dates September2012-May2016 in Erciyes University Pediatric Cardiology Department.
Results: Age of patients ranged between 4months-12months. Weight of the patients during the procedure was between 5.3-9 kg. Mean VSD diameter was 3.19 ± 0.47mm. One of defects was muscular, rest of them were perimembranous. All defects were

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closed with Amplatzer Ductal occluderII (ADO-II). Mean fluoroscopy duration and total radiation dosage were 78.5 ± 94.6 min, 2069 ± 1395 cGy/min respectively. We didnot face with any major complication except in one patient: complete AV block was seen one month after the procedure. Pacemaker was implanted. No aortic regurgitation was seen in patients after device implantation.
Conclusion: The procedure of VSD closure, whether it is surgical or percutaneous, is very risky. The risks were higher when the children were smaller than 1 year of age and low body weight. Percutaneous VSD closure may be an alternative to surgery in early infancy that carry the similar risks but less invasive.

P1465 - TRANSCATHETER EMBOLIZATION OF COLLATERAL VESSELS USING N BUTYL 2 CYANOACRYLATE IN FUNCTIONAL SINGLE VENTRICLE PATIENTS

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Background: Transcatheter coil embolization of collateral vessel such as aortopulmonary shunt (APS) and vena cava to pulmonary vein fistula is established procedure. However, it comes technically difficult when those vessels have a complex form. Also sometimes it requires many coils. As a result catheter intervention tend to be a time and money consuming procedure. N-butyl-2-cyanoacrylate (NBCA) is a monomeric acrylic glue that polymerizes rapidly when brought into contact with ionic media such as blood, and produces permanent occlusion. Utility of NBCA as an embolization material has already confirmed especially in nerouvascular area however, there are few published data that noted about its utility in congenital heart disease.
Aim: To evaluate an utility of NBCA as an embolization material for collateral vessels in functional single ventricle patients.
Case 1: Three year-old boy with Double outlet RV, MS and multiple VSD underwent TCPC at 29 months of age. We performed cardiac catheterization just before TCPC. Numerous APSs were detected by right IMA angiography. We embolized APSs with Target Coil. 360 Ultra 4mm × 6 cm was detained at distal side of right IMA. After coil detainment, we inserted microcatheter and infused 50% NBCA quickly. We confirmed complete right IMA occlusion. Case 2: Two-year-old girl with polysplenia, double outlet RV and multiple VSD. We underwent TCPS at 14 months of age. We performed cardiac catheterization just before TCPC operation. We confirmed SVC to pulmonary vein fistula. Eleven Coils were detained at distal of SVC-PV fistula. Then we inserted microcatheter to proximal side of fistula and inject NBCA quickly and fistula was completely collapsed. Arterial O2 saturation raise up from 82 to 86%
Conclusions: We achieved complete embolization of collateral vessels with coils and emulsion of NBCA in functional single ventricle patients. Embolization with NBCA may be attractive, especially for complex and small caliber vessels.

P1466 - SHORT TO MID TERM RESULTS OF TRANSCATHETER CLOSURE OF PERIMEMBRANOUS OR OUTLET TYPE VENTRICULAR SEPTAL DEFECTS USING AMPLATZER DUCT OCCLUDERS

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Objectives: We investigated the safety and efficacy of Amplatzer duct occluder (ADO) in transcatether closure of perimembranous or outlet type ventricular septal defect (VSD).
Methods: During a 4.2-year period, transcatheter closure of perimembranous or outlet type VSD using ADOs was attempted in 154 patients. Patients with a body weight <12 Kg, VSD diameter >10mm, pulmonary hypertension or moderate degree of aortic valve prolapse were excluded. Transesophageal echocardiography was used to monitor the procedure. ADOI or II were used to close perimembranous VSD, & ADOII was used to close outlet type VSDs. The size of ADO selected was 1-3 mm larger than narrowest jet width measured on the angiograms. Clinical evaluation, ECG & echocardiography follow-up were performed at 1, 3, & 6-month & annually after the procedure.
Results: Failure occurred in 7 patients including 1 complicated with ADOII migration. Of the 147 patients with successful deployment of ADOs, their age ranged from 3.3 to 65 years (20 ± 15 years). The mean Qp/Qs ratio was 1.4 ± 0.3. Of them, 125 had perimembranous type VSD & 22 had outlet type VSD. The mean defect diameter was 4.3 ±1.4 mm. ADOI was used in 74 (mean device size 6.5 ± 1.5 mm) & ADOII in 73 patients (5.7 ± 0.6 mm). No major complications occurred except 2 cases with embolization. One failure case was managed with emergent surgery. Percutaneous retrieval of an ADOI & implantation of a larger ADOI were performed in the other patient. All were available for 1 & 3-months follow-up. There were 9 patients with small residual shunt & 1 with mild-to-moderate residual shunt after a mean follow-up period of 35 ± 17 months. No one developed heart block or more than mild new-onset aortic or tricuspid regurgitation.
Conclusion: Transcatheter closure of perimembranous or outlet type VSD using ADOs is safe and effective.

P1482 - EFFICIENCY OF TRANSCATHETER PATENT FORAMEN OVALE CLOSURE WITH HYPERION PFO OCCLUDER

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Introduction: Patent Foramen Ovale (PFO) may result in paradoxical embolism event (EE). Transcatheter closure of PFO remains still interesting treatment alternative in selected patients. Experience with application of Hyperion PFO Occluders (similar to Amplatzer PFO Occluders) in such circumstancies was not evaluated yet.
Aim: To analyse clinical data and mid-term outcomes of patients (pts) with PFO after cryptogenic EE who underwent transcatheter PFO closure with Hyperion Occluder.
Methods: Among 67 patients (pts) with cryptogenic EE in whom in years 2014-2016 PFO was closed percutaneously with Hyperion PFO Occluder in our institution. There were 36 woman with mean age 41 (21-62) and 31 man with mean age 41 (19-68) years. The indications for the procedure were established by neurologist: Transient Ischemic Attack (TIA) in 15 pts, ischemic stroke in 49 pts, migraine with aura in 3 pts. Transcranial Doppler (TCD) was performed in all pts (with MES > III degree in all pts). Follow up was performed 3, 6 12 months after the procedure and thereafter yearly. Control TCD was performed after 6 months after procedure.

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Results: The procedure was successfully completed in all patients and no procedure-related complications were observed during hospitalization. Following devices were used (accordingly to the anatomy of PFO) – PFO Hyperion Occluder 18 mm (6 pts), 24 mm (49 pts), 30 mm (8 pts) and 34 mm (4 pts). Fluoroscopy time was 2,4 (0,9-22) min. During follow-up no neurological events were observed in any pt. TCD results were available in 54 pts (4 were lost from follow-up)-48 pts had complete PFO closure and 6 pts residual shunt. In 1 pt paroxysmal atrial fibrilation was observed.
Conclusion: Transcatheter PFO closure with Hyperion device is safe and effective procedure, however long-term follow-up and randomized study with other devices are necessary. ⋆Device produced as SHMSA by Lepu Company (Beijging, China)

P1484 - OFF LABEL PERCUTANEOUS PULMONARY VALVE IMPLANTATION IN SMALL CONDUITS

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Background: Guidelines allow percutaneous pulmonary valve implantation (PPVI) in conduits above 16 mm diameter. Balloon dilatation of a conduit to a diameter greater than 110% of the original implant size is also not recommended. We analyzed patients undergoing PPVI in such conditions.
Materials and Methods: Nine patients (May 2008-July 2016) from 8 institutions underwent PPVI in conduits < 16 mm diameter. Five patients with 16-18 mm conduit diameter underwent PPVI after over-expansion of the conduit >110%. Mean age and weight of the 14 patients was 12.1(7.7 to 16) years and 44.9(19 to 83) Kgs.
Results: Median conduit diameter at PPVI was 12(10 to 17) mm. Median systolic right ventricular pressure was 70(40 to 94)mmHg. Procedure was successful in all cases. A confined conduit rupture occurred in 7 patients (50%) and was treated with covered stent in 6. One patient experienced dislocation of 2 pulmonary artery stents that were parked distally. The post-implantation median systolic right ventricular pressure was 36(28 to 51) mmHg. A fistula between right-ventricle outflow and aorta was found in one patient, secondary to undiagnosed conduit rupture. This was closed surgically. After a median follow-up of 27.4(2.5 to 93.8) months, all the patients are asymptomatic with no significant RVOT stenosis.
Conclusions: PPVI is feasible in small conduits but rate of ruptures is high. However such ruptures remain contained in the majority of cases because of surrounding fibrosis. It is then possible to implant the largest available pulmonary valve in small conduits. Surgical back-up should be available in case of uncontrolled conduit rupture.

P1488 - ATRIAL SEPTAL STENT IMPLANTATION FOR THE MANAGEMENT OF CONGENITAL HEART DISEASE IN INFANTS

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Introduction: The creation of an unrestrictive atrial septal defect could be crucial for the management of complex congenital heart defects in different situations, such to relieve atrial hypertension, to maintain cardiac output, or to achieve adequate atrial mixing to improve systemic oxygen saturation. The aim of this study is to report an institutional experience performing percutaneous atrial septal stenting to relief atrial restrictions in various scenarios.
Objective: We retrospectively reviewed all pediatric cardiac catheterization procedures that included transcatheter atrial septal stent implantation (or an attempt), that were performed between 2005 and 2015.
Results: Thirty-seven patients underwent 40 transcatheter interventions. Thirty-nine implantations attempts (97,5%) were successful. The median weight was 4 kg [2-80 kg] and median age 3,5 months [0-193 months]. About 40% of patients had an intact atrial septum (IAS), and radiofrequency perforation of the interatrial septum was used before septal stent implantation. Left or right venous femoral access, were used in all patients. Indications were categorized in: to relief left atrial hypertension (45%) in the setting of hypoplastic left heart syndrome (N = 14) or left heart obstructive defects (N = 4); to maintain cardiac output in the setting of severe pulmonary hypertension or right ventricle dysfunction (35%); and to unload left cavities in the context of severe left ventricular dysfunction and ECMO assistance (20%). Major adverse events were seen in 12% of procedures: 3 stent migration, and 2 cardiac tamponade (one periprocedural death). About 20% of patients required stent overdilation. All stented atrial septum remained widely patent until elective surgical stent explant (54%), or until last follow-up (median follow-up time 32 months).
Conclusion: A restrictive atrial septal defect or IAS, could be a challenging problem in different situations, for the management of cardiac diseases in infants. Atrial septal stent implantation could be a feasible and effective technique in selected situations.

P1490 - FIRST 100 NEW COBALT CHROMIUM STENTS IMPLANTED IN LARGE VESSELS AND OTHER POSITIONS

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Background: Stent implantation is well established method of treatment in patients with large vessels narrowing. We present our experience with application of first 100 new cobalt – chromium AS XL/XXL stents implanted in several positions.
Materials and Methods: There were 98 patients treated with 100 stents - 57 (aged 8-65 years) with native aortic coarctation (CoAo) or recoarctation, 17 (aged 6-64 years)with left or right or left PS closely to the bifurcation (native or postsurgical). In 21 pts (aged 11-40 years) the procedure was presenting before Melody valve implantation (in calcified pulmonary homograft or native right ventricle outflow tract- RVOT). In 3 patients stents were implanted in some other places to dilate stenosis of: superior vena cava (in 7,5 years old child), Fontan tunnel (in 17 years old boy), and atrial septal defect (interatrial septum in complex heart defect in 19 years old boy). Mean follow-up was 3,4 (0,2-5,4) years.
Results: All procedures were finished successfully in all but two patients without any complications with significant clinical improvement. Two migration of stents occurred – one in RVOT and another in left pulmonary artery (without clinical consequences). In all cases successful dilation of stenosed place with

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significant gradient reduction occurred. In 2 cases of native CoA (23 and 34 years old man) in early follow-up (6 and 8 months after the procedure) in angio CT small aneurysm formations was observed. Both patients were treated successfully with covered stents. In follow-up in one patient (presented for valve implantation) stent fracture was found and no other complications were observed.
Conclusions: Implantation of new cobalt-chromium AS XL and XXL is a good therapeutical option for the treatment of stenosed great vessels.

P1556 - PATIENTS AGE IMPACT ON THE RESULTS OF THE TRANSCATHETER ATRIAL SEPTAL DEFECT CLOSURE

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Transcatheter closure with self-expandable double disc devices (DDD) became a method of choice in the treatment of secundum atrial septal defects (ASD). The complications remain the main concern of the procedure despite the gained experience. The aim of the study is to assess the relationship between the patients age and event-free procedure.
Patients and Methods: We retrospectively reviewed the data base of 169 consecutive patients, 112 children, mean age 9,8 (3,2) years (group 1) and 57 adults, mean age 39 (14,6) years (group 2), with attempted DDD, Amplatzer type, performed in a tertiary heart center.
Results: Event-free course was observed in 148 pts. (88%). There were 6 major complications (1 death due to unrecognized retroperitoneal bleeding; 3 early and 1 late embolizations with surgical removal and 1 acute pulmonary edema in patient on chroniodyalisis). Fifteen minor complications were observed (3 explantations before release due to unstable DDD position; 3 transcatheterly removed embolizations; 1 gastrointestinal bleeding; 1 small pericardial effusion; 7 postprocedural dysrhythmias - atrial fibrillation/flutter). Table. Comparison of the type and rate of complications in both groups
Conclusions: ASD device closure is an effective and relatively safe procedure at any age. Patients age has no impact on the major complications and embolizations. They are related mainly to the preprocedural assessment of the ASD size and morphology. Patients age influences dysrhythmias that need precise pre- and postprocedural estimation and treatment.

Table.

	Group 1(N = 112)	Group 2(N = 57)	p
Closure rate (%)	97	92	ns
Total complications (%)	4,6	28,6	0,000
Major complications (%)	1,9	7,1	ns
Total embolizations (%)	2,8	7,1	ns
Dysrrhythmias (%)	0,9	10,7	0,007
TTE size (mm)	14,3 ± 3,1	18,6 ± 4,4	0,000
TEE size (mm)	14,8 ± 3,3	20 ± 5	0,000
Qp/Qs	1,9 ± 0,5	2,2 ± 1	ns
Device size (mm)	16,5 ± 4	23,3 ± 5,7	0,000
Difference DDD-TTE (mm)	2,1 ± 2,4	4,7 ± 4,2	0,000
Septal aneurism (%)	35,2	41,1	ns

P1559 - RIGHT VENTRICLE DEPENDENT CORONARY CIRCULATION IN A NEWBORN CRITICAL PULMONARY VALVE STENOSIS

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Introduction: Coronary-cavitary connections often associated with pulmonary atresia with intact interventricular septum, may occur in neonates with critical pulmonary stenosis (PS), which adds a worse prognostic.
Objectives: To describe a rare case of critical PS with coronary circulation dependent on the right ventricle (RV) .
Case report: –baby girl, born at term, BW = 3350 g. Maternal history of gestational diabetes and systemic arterial hypertension. A systolic murmur was heard at the first day, being well in hospital time she was discharged home on the third day of life. On her ninth day, she presented to a pediatric cardiologist with tachycardia and weight loss. In her investigation: Chest x-ray showed mild cardiomegaly, slight oligaemic pulmonary fields ; ECG - sinus rhythm, QRS + 60° and incomplete RBBB. Echocardiographic findings: critical PS, PFO with bidirectional shunt, moderate tricuspid regurgitation, a hypertrophied good-sized right ventricle (RV). Estimated RV pressure was 118 mmHg, and a 2mm patent ductus arteriosus. She was admitted to the hospital and started on prostaglandin. At 28 days of life, she was referred to our center for pulmonary balloon valvuloplasty (PVB). Hemodynamics findings: RV-PA gradient of 45 mmHg and RV/AO ratio of 1.39. After RV angiography, PVB with a Power-flex balloon 10 × 2, was performed, followed its deflation, the patient went to complete atrioventricular block, cardiogenic shock and death despite exhaustive CPR manoeuvers. Going back to review her angiographies the left coronary artery were seen arising directly from the right ventricular outflow tract.
Conclusion: decompression of the RV was probably the cause of death. Aortography should be performed prior to PVB in suspected cases

P1569 - LONG TERM RESULTS OF ULTRAHIGH PRESSURE BALLOON ANGIOPLASTY FOR PERIFERAL PULMONARY ARTERY STENOSIS. COMPARISON ANALYSIS WITH STENT PLACEMENT

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Background and Objectives: Balloon angioplasty for postoperative pulmonary artery stenosis is an important therapeutic option to maintain and facilitate the pulmonary circulation. During the past decade, manykinds of non-compliant balloons which provide excellent trackablity have been widely used. Especially, efficacy of ultra-high-pressure balloons (UHPB) such as CONQUEST@ has been also reported, but their long term benefits are not known. To analyze the long term efficacy of UHPBs against postoperative branch pulmonary artery stenosis compared to stent implantation.
Methods: Retrospective analysis of follow-up catheterization data over three years after angioplasty by UHPBs or stents against the lesions with biventricular corrective surgery. Five cases, 11 branches of UHPB group and 8 cases, 9 branches of stent groups are included.

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Results: Mean follow up period was 4.7 years in UHPB group, and 6.8 years in stent group. Mean gain of minimal lumen diameter (MLD) at post-/pre- angioplasty was 122 (100-300) % in UHPB group and 189 (131-560) % in stent group. Whereas the changes in MLDs between the follow up period (MLD at the latest follow up/just after initial intervention) was 162 (129-231) % in UHPB group and 102 (63-143) % in the stent group, which displays significant advantage in vascular growth in UHPB group ($P < 0.01$). Balloon redilation procedure was performed 0.8 times in UHPB group and 3.0 times in stent group.
Discussion and Conclusions: Stent implantation revealed efficacy in acute vascular dilation and its maintenance. Furthermore, UHPB provided long-term benefits in vascular growth.

P1570 - TWO CASES OF COMPLICATION OF CORONARY FISTULA AFTER AMPLATZER SEPTAL OCCLUDER

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Background: Transcatheter occlusion of atrial septal defect (TC-ASD) has been first treatment option of this disease, but various complications such as migration and embolization, erosion have been reported.
Purpose: We report another rare complication of TC-ASD, development of multiple coronary artery fistulae found in two adult patients. Numerous fistulous coronary arteries run toward inter-atrial septum and surround device.
Cases: Patient 1: 43-year-old female underwent successful closure of ASD; 26 mm in diameter, using 30mm Amplatzer septal occluder. She did not have coronary artery disease or coronary fistula. When she underwent transesophageal echocardiography (TEE) one year after TC-ASD as our protocol, turbulent flows were demonstrated around the device. Cardiac catheterization revealed numerous coronary fistulae running from both left and right coronary arteries toward the device giving Qp/Qs ratio of 1 to 1.42. Patient 2: 72-year-old female underwent successful closure of ASD; 24.2 mm in diameter, using 26mm Amplatzer septal occluder. Because she was complicated by pulmonary hypertension, she underwent cardiac catheterization periodically that did not showed coronary abnormality up to 16months after TC-ASD. After 4 years and 8 months, we found numerous coronary fistulae running from both left and right coronary arteries towards the device.
Conclusions: We report development of coronary fistulae as rare complication after TC-ASD. Cardiologists must be aware of this complication and needs careful follow up of these patients. Obviously, further study is required to determine frequency and clinical implication of this complication.

P1585 - AORTOARTERITS MYRIAD MANIFESTATIONS CHALLENGING DECISIONS SHORT AND MEDIUM TERM OUTCOMES

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Introduction: Aortoarteritis is a chronic inflammatory disorder of aorta and branches. The etiology is unknown, disease progression with stenosis/aneurysm formation is difficult to monitor and treatment is not standardised.
Methods: Retrospective and prospective. From January 2011 to January 2017 all children with takayasu arteritis at a tertiary care pediatric cardiology centre.
Results: Total of 33 children with takayasu arteritis were seen. Median age was 12 years (range 3 yr-18yr). M: F was 1:3. Hypertension 32(96%) and cardiac failure 27(81%) were the common presentations. Elevated ESR (>20) was seen in 38% (10/26). Renal artery stenosis with shrunken kidney was present in 11 (33%) children at presentation including a 3 year old(R -4, L -6, b/l- 1). 21 (64%)children (R-2, L-7, b/l 11,) had renal involvement and 21(64%) had aortic involvement. Aortic arch branches were occluded in 12(36%). Biventricular dysfunction was seen in 4 patients suggestive of myocarditis. Thirteen children underwent aortic interventions (balloon 6, stenting 7). Thirteen children underwent renal intervention (Balloon 6, Stent 7). There was normalisation of blood pressure and resolution of failure after intervention. Methotrexate and steroid were used for immunosuppression. Aspirin was given to all stented children. Seven children had evidence of tuberculosis. Three children are lost to

Table.

	Sex	Age	Presentation	Involvement	Intervention	F/U	ESR	Reintervention	Treatment
1.	F	11	F, CCF	LR	US	0	0		
2	M	9	F, CCF, HTN, TB	BR	US	12	28		WL,ATT
3	F	8	F, HTN, CCF, AHTN, APE	BR, AbA	R,L, AbA,B	24	38		MP,WL, ATT, MTX
4	F	19	HTN	LR	US	12	0		WL,ATT, MTX
5	F	11	F, HTN, BVF	BR, AB, F	R,L,S	6-D	10		MP,MTX
6	F	13	F, CCF, HTN	AbA	A,S	48	40		WL,ATT
7	F	11	F, CCF,HTN	BR, AbA,	R,L B	36	40		WL,ATT, MTX
8	F	9	F, CCF, HTN, APE	RR	R,S	24	6		WL,ATT
9	M	14	F, FTT, CCF, HTN	BR	L,S	18	28	R	WL,MTX
10	M	12	HTN	AbA, AB	A, B	0	0		0
11	M	4	F	CF,coronary, LR,AbA	R,B	1	70		MP,IVIG
12	F	12	F , CCF	LR, AbA	US	0	0		0
13	M	17	HTN	LR	US	20	6		ANTI HTN
14	M	13	HTN	TA	TA ,S	D			DIED
15	F	14	F, HTN,APE	AbA	AbA,S	D	10		MP,MTX
16	F	12	F, CCF, TB, BVF	AbA	AbA,S	22	20		WL
17	F	8	HTN, CCF	LR, AbA	US	18	29		MTX
18	M	5	F,FTT	AbA, TA	AS	24	44		MTX
19	F	17	F, CCF,APE	LR,TA,AB	R SS	10	20		MP,MTX
20	F	3	F, CCF,FTT, APE,TB	BR, AA,TA, AB	R B	6	20	R	WL,MTX
21	F	16	F, claudication, MR	AB, CELIAC MILD	SC B	7	16		WL,MTX
22	F	14	F, CCF, HTN	TA	TA,S	D	58		Warfarin, MP,WL, MTX
23	F	16	F, CCF,HTN	BR,AB	RS	13	20	R	WL,MTX
24	M	13	F,CCF,HTN	AbA	AbA,S	13	10	R	WL,MTX
25	F	12	F, APE,CCF	BR	US	14	10		MTX
26	F	9	F, CCF, APE, HTN,BVF	BR, AbA	R L S	10	5		WL,MTX
27	F	10	F, CCF,APE, HTN	BR	US	19	12		WL,MTX
28	F	13	F,HTN	AbA	AbA B	0	0		WL,MTX
29	F	7	F, TB,BVF	TA	TA B	3	10		WL,MTX, ATT
30	F	18	HTN	TA, AbA	TA S/AbA B	4	40	R	MMF,WL, MTX
31	F	17	F	BR, AbA	R B	10	20		WL,MTX
32	F	17	F, HTN	AbA	AbA SB	36	8	R	MTX
33	M	17	HTN	RR	R B	3	10		MMF, MTX

Abbrevations: F- Fever: CCF – congestive cardiac failure: FTT – failure to thrive: APE –acute pulmonary edema: HTN-hypertension: TB- tuberculosis: MR-mitral regurgitation: BVF-biventricular failure: LR-left renal: RR-right renal: BR-bilateral renal: AB- aortic branches: AA-ascending aorta: TA-thoracic aorta: AbA-abdominal aorta: CF-common femoral: US-unsuccesful: R-renal:A-aorta:B-Balloon:S-stenting: D-dead:Mp-methyprednisolone:WL-wysolone: MTX-methotrexate:MMF-mycophenolate mofetil: ATT-antitubercular therapy

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follow up. Follow up ranged from 3 to 48 months. Six (20%) children required re intervention. Four (13%) children died, three in acute phase and one after 6 months.
Conclusion: Hypertension and heart failure are most common manifestation. End organ damage can be present at initial presentation. Monitoring disease activity is difficult. Intervention with ballooning + stenting is lifesaving. There is a strong association with tuberculosis. Steroids and methotrexate are not effective in preventing recurrence. Studies with newer anti-inflammatory drugs are necessary.

P1586 - 3D PRINTING TECHNIQUES FOR INTERVENTIONAL PLANNING

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Background: The spatial relationships of anatomical structures in patients with complex congenital heart disease can represent a challenge for interventional planning. Cardiac magnetic resonance and multi-detector computed tomography provide information that can be reproduced in a 3D printed model to assess the feasibility of interventional treatment.
Methods: Interventional planning was reproduced using a patient-specific 3D-printed heart in cases in whom cardiac catheterisation was controvertible. Segmentation was performed from previous imaging acquired dataset (CMR/MDCT) with Mimics software. Models were fabricated by polyjet technology or fused deposition modelling. Cardiac catheterisation was performed in the model before proceeding in the patient.
Results: Seventeen patients were referred for cardiac 3D-printing (88% adults). Five patients had partial anomalous pulmonary venous drainage(PAPVD); three transposition of the great arteries(TGA); five coronary artery fistulas, one aortic aneurysm post aortic valve replacement; one tetralogy of Fallot (TOF) and one had multiple ventricular septal defects (VSD). 65% patients underwent cardiac catheterisation, one coronary fistula and one TGA (13%) were considered unsuitable after reviewing the model and 35% are awaiting further management. LPA stenosis motivated printing for 2/3 of TGA patients and the TOF case, stent length and size were selected based on the models. The remaining TGA had a previous Senning operation, developing a baffle leak that was considered unsuitable for stent closure given its proximity to the mitral valve. Three coronary fistulas were successfully closed. Occlusion of the aortic aneurysm was achieved using an AVP II device and a large VSD was closed with an amplatzer device in the case with multiple VSDs. All patients who underwent catheterisation had a successful result.
Conclusions: 3D models provide the opportunity to select materials and plan access routes before cardiac catheterisation. This is beneficial where spatial complexity obstructs expert led treatment, providing the interventional cardiologist with confidence to address conditions that might otherwise be resolved with cardiac surgery.

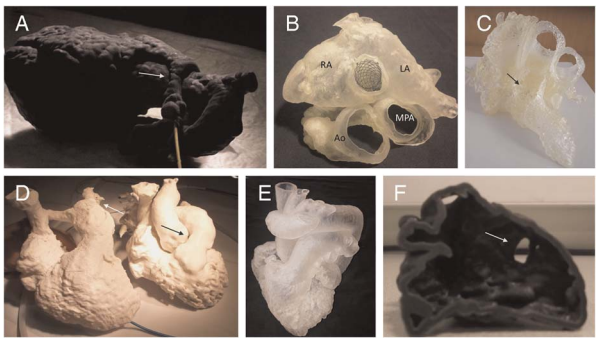

Figure 1.
A) Left pulmonary artery (LPA) stenosis after Le Compte manoeuvre, balloon and stent sizing pre – procedure. B) Baffle leak after atrial switch operation. Visualization and planning of stent route in the superior vena cava (SVC), RA=Right atrium, LA=left atrium, Ao=aorta, MPA=main pulmonary artery. C) Aortic aneurysm post aortic valve implantation (arrow). D) LPA stenosis (white arrow, left hand side) and RVOT stenosis (black arrow, right hand side). E) 3D model for patient with partial pulmonary venous drainage (PAPVD). F) Interventional closure of large ventricular septal defect (VSD) (white arrow) in patient with multiple VSDs.

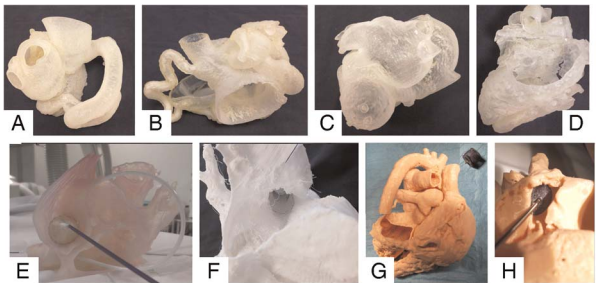

Figure 2.
A) Right coronary artery (RCA) fistula to left ventricle (LV). B) Coronary fistula involving both right and left coronary arteries. C) Complex coronary artery fistula with two aneurysms. D) Left main stem coronary fistula draining in the superior in the vena cava (SVC) – right atrium (RA) junction. E&F) PAPVD: SVC stent redirecting right upper pulmonary vein flow to left atrium. G&H) Procedure planning for left main coronary fistula draining into SVC to RA junction.

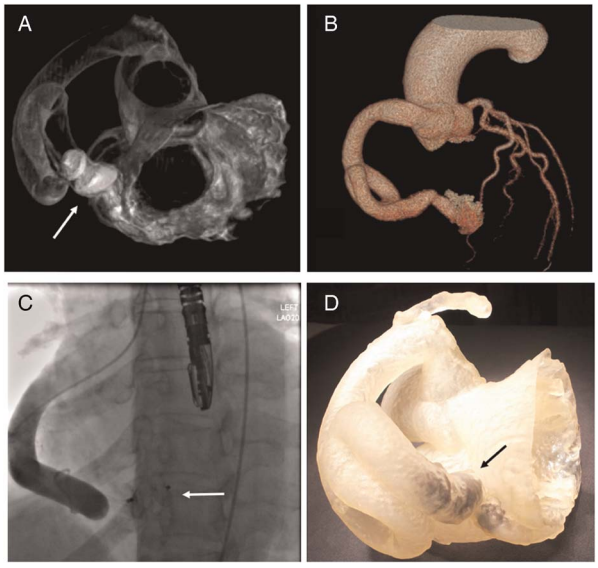

Figure 3.
Example of procedure planning and intervention using 3D printing techniques in a patient with RCA to LV fistula. A) Rotational angiography of the model with device at drainage site. B) Volume - rendered image from CT images showing the fistula. C) Post-release angiography showing amplatzer in situ with no residual flow. D) 3D model with device as shown in figure A.

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P1590 - ECHOCARDIOGRAPHIC FEATURES OF SECUNDUM ATRIAL SEPTAL DEFECTS ASSESSED AS UNSUITABLE FOR TRANSCATHETER DEVICE CLOSURE. A PEDIATRIC SINGLE CENTER EXPERIENCE

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Objectives: To review our experience about morphological characteristics of secundum atrial septal defects (ASDs) for which percutaneous closure was thought inappropriate.
Methods: The study included 46 consecutive patients (mean age 7.67 ± 3.9 years; range: 2-1 years) assessed as unsuitable for transcatheter device closure of an secundum ASD, either directly referred to surgery or after assessment in the catheter laboratory between 2012 and 2016. Patients data, echocardiographic findings and procedural informations were retrospectively reviewed from the medical records.
Results: One hundred and fifty-one children underwent closure of an secundum ASD in our institution between 2012 and 2016 either surgically (n:46, 30%) or percutaneously (n:105, 70%). Among the patients with surgically closed, 33 children was referred directly to the surgery, 7 were thought unsuitable for device closure during the catheterization and 6 were referred after failed transcatheter attempts. Patients were referred to the surgeon for the following reasons: defects with deficient rim(s) (n:23), insufficent total septum(8), defect sized too large(n:6), multiple defects (n:4), aneurysmal IAS (2), a defect close to the superior vena cava (n:2), device entrapment within a Chiari network (1).
Conclusion: Transcatheter closure of isolated secundum ASD provides, excellent results with a lower complication rate with a shorter hospitalization period. However some of ASDs are not suitable for a percutaneous closure. In accordance with the current litaratures rim deficiencies are the most common reason of deferral of device closure was found in our study. However the requirements of large implants, defiencies of total septum, presence of multiple defects and some of other causes continue to constitute an impediment in front of the percutaneous closure.

P1619 - THE ASSESSMENT OF THE EFFECTIVENESS OF BALLOON ANGIOPLASTY OF NATIVE AORTIC COARCTATION IN NEWBORNS AND INFANTS YOUNGER THAN FIVE MONTHS OWN EXPERIENCE

Jacek Kusa[1], Pawel Czesniewicz[2], Agnieszka Skierska[3], Leslaw Szydlowski[3], Malgorzata Ras[2], Elzbieta Skiba[2], Elzbieta Wojcik[2]
Medical University of Sielsia/Regional Specialized Hospital, Pediatric Cardiology Department, Wroclaw/Katowice-Poland[1]; Regional Specialized Hospital, Pediatric Cardiology Department, Wroclaw-Poland[2]; Medical University of Sielsia, Pediatric Cardiology Department, Katowice-Poland[3]

Introduction: Balloon angioplasty of aortic coarctation in children is an established method of treatment but is not recommended in patients under six months of age due to significant risk of rapid recoarctation. The aim of study is to evaluate the effectiveness of this method in newborns and youngest infants.
Material/Methods: Between 01.03.2011 and 01.12.2016 balloon angioplasties of aortic coarctation were performed in 21 patients aged between 6 and 149 days (mean-80 ± 56). Only patients with naïve form of coarctation were qualified for percutaneous procedure. There were 7 newborns (33.3%). Eight patients had a bicuspid aortic valve. Seven patients (33.3%) had critical form of coarctation (heart failure), five (24%)- patent ductus arteriosus (2 of them with Prostin).
Results: After procedure the stenosis diameter increased from 1.9 (0,4-2,8) to 4.1mm (1,9-6,2)mm. The maximum echocardiographic gradient decreased from 70.6 (48-120)mmHg to 28,2 (15-38)mmHg, average from 37 (21-62)mmHg to 11,4 (6-17)mmHg, and hemodynamic gradient from 35.7 (6-69) to 9.3 mmHg (1-30)mm Hg. There were complications associated with procedure- limb ischemia in two cases (9,5%), In one- the normal blood flow returned after heparin administration. In the second patient, despite heparin and actylise the pulse did not returned, but the efficient collateral circulation developed. During FU- 39 ± 22 (6-69) months 6 patients required some intervention. Three patients (14,3%) required surgical plasty (1 month after angioplasty) because of the ineffectiveness of percutaneous intervention, in another 3 patients the treatment was effective, but during FU stenosis returned and after 18 (6-36) months the successful repeated balloon angioplasty was performed. In the remaining patients the good effect of treatment is still present.
Conclusions: Percutaneous angioplasty of aortic coarctation in the youngest age groups may be an effective treatment, alternative for cardiac surgery. The incidence of recoarctation during medium-term FU does not differ significantly from cardiac surgery and can be successfully treated by repeated percutaneous angioplasty.

P1621 - ASYMPTOMATIC EARLY NON INFECTIOUS THROMBOSIS OF THE MELODY® VALVE

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Bioprosthetic valve thrombosis is an uncommon complication. Acute dysfunction of a bioprosthetic valve may lead to hemodynamic compromise and evaluation with initiation of appropriate therapy is required. We report a patient with prior uncomplicated percutaneous pulmonary valve replacement (The Melody®, Medtronic, Inc. Minneapolis, MN) who later developed early asymptomatic severe pulmonary valve stenosis without evidence of infection. A 17-year- old male with congenital ventricular septal defect and pulmonary atresia underwent uncomplicated transcatheter pulmonary valve replacement with a 22-mm Melody® valve. At 4 months post-implant echocardiography revealed 80 mmHg gradient at the Melody® valve. Stent integrity was normal. Two blood cultures were obtained independently and were negative. Thrombophilia risk factors was normal. His D-dimer was found elevated at 1.1 mg/L (≤ 0.55 mg/L). After recombinant tissue plasminogen activator (rt-PA) infusion gradient decreased. Repeat echocardiogram demonstrated normal appearance of the Melody® valve leaflets and gradient of 28 mmHg. Acute changes in Melody® valve function should prompt a thorough investigation for and treatment of thrombosis.

P1668 - SAFETY AND EFFICACY OF TRANS CATHETER CLOSURE OF SMALL RESTRICTIVE PERIMEMBRANOUS VSDS WITH MIDTERM FOLLOW UP IS IT REALLY SMALL

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Introduction: Transcatheter closure of small to medium perimembranous VSD is gaining its importance with less traumatic technique. In a developing country like India we found many reasons to close such VSDs like: 1) Recurrent respiratory tract infections (RTI) and poor weight gain, 2) Denial of pediatrician to treat simple URTI, 3) Financial burden to treat the morbidities, 3) psychological trauma to the family and patient inspite of counseling, 4) Social issues like denial of job and marriage.
Materials and Methods: A prospective study was designed September 2014 – October 2016. A total of 254 cases of VSD device closures were done using ADO II devices. Age ranged from 0.5 to 18 years (median 5 years), weight ranged from 5.0 to 55 kg (median 14 kg), height ranged from 59 to 169 cm (median 105 cm), Median fluoroscopic time was 6.25 minutes (1.27 to 58.16 minutes) and the median VSD diameter was 4mm (3 to 6 mm).
Results: The complete VSD closure rate was 93%, 95% and 97% at 6 months, 1 year and 2 years follow-up respectively. At 6 months follow up most children demonstrated appropriate weight gain reaching weights >10th centile, decreased incidence of RTI. Immediate complications were one device embolization to the pulmonary artery, another patient had severe aortic regurgitation (AR), both devices were retrieved successfully and the patients were sent for surgery. One patient required PPI for complete AV dissociation, which later on reverted to sinus rhythm. 15 children developed new onset trivial AR which remained same at 2 years follow up.
Conclusion: Our study clearly demonstrated beneficial effects of transcatheter closure of small VSDs especially with the newer ADO II device in terms of improved left ventricular dimensions, better weight gain and freedom from recurrent respiratory tract infections in small children with negligible midterm complications.

P1676 - AN UNUSUAL CAUSE OF CHEST PAIN AFTER PERCUTANEOUS CLOSURE OF ATRIAL SEPTAL DEFECT IATROGENIC PNEUMOMEDIASTINUM

Timur Meşe[1], Murat Muhtar Yilmazer[1], Aysun Hacer Saritaş, Rahmi Özdemir[3]
Izmir Dr.behçet Uz Children, Pediatric Cardiology, Izmir-Turkey[1]; Izmir Dr.Behçet Uz Children, Pediatrics, Izmir-Turkey[2]; Dumlupinar University Evliya Çelebi Training and Research Hospital, Pediatric Cardiology, Kütahya-Turkey[3]

Introduction: We present an unusual complication associated with ASD closure procedure .
Case: A seven years old boy with 11 mm secundum ASD was electively scheduled for percutaneous ASD device closure. His preprocedural examinations were normal. In our instution anestiesiology clinic prefers to use deep sedation of a pediatric patient with ketamine during the procedure. In this case anetesiologist switched to genereal anestesia with intubation because of an ineffective sedation. Standart percutaneous ASD device closure procedure was completed without any problem or complication with the guidance of transesophageal echocardiography (TEE). After completion of procedure some blood in the saliva was seen on the TEE probe. Examination of oral cavity and throat revealed a 3-4 mm mucosal laseration in the retrofarengeal area without any persisting bleeding. Post procedural period under monitorization was uneventful up to 4 hours. When child became fully concious he had complained a chest pain. Vital signs and standart 12-lead-ECG was normal. An urgent echocardiographic examination was unsuccessful because of an insufficient visualization of the cardiac structures in many echocardiographic windows. His chest x -ray revealed a lienear air lucency on the border of left ventricle suggested the diagnosis of pneumomediastinum strongly, which was confirmed with lateral chest x ray. This "air curtain" between our probe and heart prevented to get an optimal echocardiographic images. Patient followed up for 3 days there was no sign or clue of any mediastinitis, cardiac or respiratory distress. After three days of follow up pneumomediastinum disappeared spontaneously and echocardiographic images were fully obtained.
Conclusions: Pharyngeal abrasions and hemorrhage are the most common complications associated with TEE insertion. Esophageal perforation, a rare but life-threatening event can cause hemorrhage, subcutaneous emphysema and pneumomediastinum.

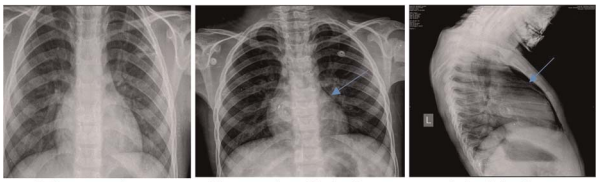

Figure.

P1688 - MID TERM FOLLOW UP RESULTS OF THE SOLYSAFE SEPTAL OCCLUDER FOR PERCUTANEOUS CLOSURE OF SECUNDUM ATRIAL SEPTAL DEFECTS

Murat Muhtar Yilmazer[1], Selman Vefa Yildirim[2], Timur Meşe[1], Bariş Güven[3], Savaş Demirpençe[1], Rahmi Özdemir[4], Taliha Öner[1], Vedide Tavli[1]
Izmir Dr.Behçet Uz Children, Pediatric Cardiology, Izmir-Turkey[1]; Near East University, Pediatric Cardiology, Nicosia-Cyprus[2]; Tepecik Training and Research Hospital, Pediatric Cardiology, Izmir-Turkey[3]; Dumlupinar University, Evliya Çelebi Training and Research Hospital, Pediatric Cardiology, Izmir-Turkey[4]

Objective: Septal occluder (SSO) (Swissimplant AG,Solothurn, Switzerland) which was withdrawn from the market, due to wire fractures since 2010. Limited data exists on the Solysafe device and outcomes related to this ASD device closure in children. We report mid-term results of the patients whose defects closed percutaneously with SSO.
Methods: A total of 25 patients– with mean age of 8.44 ± 3.6 years with secundum atrial septal defect (ASD) underwent attempted transcatheter closure between July 2008 and June 2010. The device was implanted successfully in 22 of 25 patients. Mean follow up period was 6.10 ± 0.53 years.
Results: Mean stretched diameter of the ASD determined by balloon sizing was 13.6 ± 4.43 mm. Nine 15-mm devices, eight 20-mm devices, six 25-mm devices and two 35 mm devices were used. A twenty mm, and 2 of 35 mm devices were intended to be used in 3 patients whose attempts were unsuccessful. There were 2 major complications (8%). First, an embolization into the pulmonary artery in a 17-year-old girl. Another case was a 11 year-old girl who had implantation of 15 mm SSO device into the 8 mm defect was developed right hemiparesis after the procedure. During the 6 years of follow-up we did not observe wire fractures, pericardial effusion, endocarditis, hemolysis, ECG changes, valvular problems or suspicious echocardiographic findings. We observed device embolization and hemiparesis as major complication related to the procedure
Conclusions: We did not observe any significant complication in the mid term follow-up period excluding major complications during periprocedural period. Probably the main reason not to observe wire fracture might be related the selection of appropriate

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patient with smaller defect size. Although the device was fully covered by endothelial tissue, long term monitoring must be requried for wire fractures and related complications.

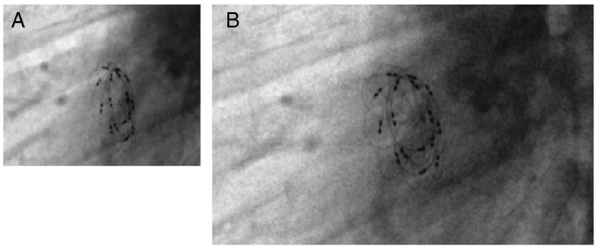

Figure 1.

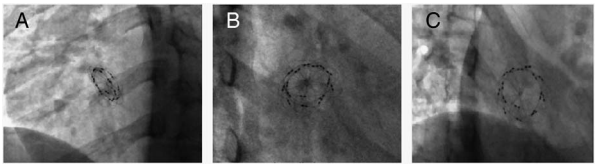

Figure 2.

P1697 - ENDOVASCULAR VSD CLOSURE

Jesus Damsky Barbosa[1], *Fernanda Biancolini*[2], *Julio Biancolini*[3], *Adelia Marques, Victorio Lucini*[4], *Jaime Cornejo*[4], *Judith Ackerman*[5], *Ana De Dios*[3]
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Objective: to demonstrate that the endovascular treatment is an alternative for VSD closure.
Material and Methods: Observational analysis. From May 2010 to March 2016. 47 patients (p) were treated. Median age: 7 yo (0.33 to 15a). Median weight: 26 kg (4.3 to 83 kg); 5 p <10 kg. Sex: 28 f 19 m. Complications: Major: severe. Minor: mild. Devices: PFM® coils, Amplatzer devices, CERA® (Lifetech) and MEMOPART® (Lepu Medical Technologie).
Results: Successful: 43 procedures (84.3%). Frustrated: 4 procedures: 2 AV Block. 1 embolization (musc Cera). 1 p: acute collapse of the right ventricular. Types: 20 perimemb VSDs (46.5%), 11 high musc (25.5%), 6 midventricular (13.9%), 3 apical (6.9%) and 3 postsurgical (6.9%). Complications: Major: 3p (6.9%): 1 tricuspid stenosis and 1 hemolysis (PFM); 1 p <10 kg post-procedural severe AR (ADO II) followed by death. Minor: 3 arrhythmias: required steroids (2: Lifetech, 1: ADO II); 1 p: the device embolized and repositioned (Lifetech); 2 p: transient hematuria (PFM); 1 coil fracture (PFM); 2 p: moderate tricuspid insufficiency (PFM); 1 patient: mild to moderate aortic insufficiency (PFM). The most important complications were during our firsts cases experience. Devices: Le VSD coil: 3 hemolysis: 2 self-limited, 1 severe. 1p tricuspid stenosis that required surgery. 1 fracture a week after procedure. Cera 12 p: 2 embolizations within the first cases, 1 residual shunt treated at 2 years (residual shunt vs other VSD). Memopart: 8 p. We had no complications or residual shunt. Amplatzer: 7 p: 3p ADO II. 4 p muscle VSD. There was no AV block at follow-up.
Conclusions: 1) The endovascular VSD closure is a feasible procedure. 2) The existence of different types of devices allows closing different types and sizes of VSDs. 3) There were not AV block along the follow up. 4) The learning curve was inexorable.

P1699 - FONTAN CIRCULATION CATHETER INTERVENTIONS AND MORTALITY

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Introduction: Patients with Fontan circulation demand a careful follow -up. Cardiac interventional catheterizations are an essential tool in the management of these patients .(pts)
Objective: The aim of this study was to describe the clinical indications of percutaneous interventions and related mortality in Fontan's pts.
Methods: Between 1989-2016, 431pts underwent a modified Fontan procedure with a median follow-up of 5,6 years (IQR--75 = 2,25-11,1 years). In 113pts, 159 catheter interventions were undertaken. According to the median interval between surgery and the first interventional catheterization two groups were identified: Group I: early: 0.05 years(IQR 0.04-0.1), 27pts. Group II: late 2.07 years(IQR 1.2-4.1), 86 patients.
Results: The most frequent clinical indication in group I was a Fontan circulation dysfunction (p 0.0000) and an increased cyanosis in group II (p0.0000). Hemoptysis was the main event in those patients requiring subsequent procedures (p0.0001) The percutaneous treatment included: group I: balloon fenestration dilation 8p(29,6%), balloon dilation of pulmonary branches 9p (33,3%) stent deployment in 7p(25,9%), and thrombolysis 3p (11.1%). group II: fenestrations closure 42p(48,8%) and coiling aortopulmonary collaterals 37p(44,2%). There were 13 deaths (11.5%): group I : 9pts(32,1%) group II : 4pts. (4,7%), (p 0.0002). By univariate analysis mortality was associated to Fontan circulation dysfunction (p0.0000), early postoperative period (p0.0000), balloon fenestration dilation (p0.003)and increased cyanosis (p0.04). Multivariare analysis mortality was related to Fontan circulation dysfunction OR 11,69 (1.73-79.09) p 0,01
Conclusions: Fontan circulation dysfunction was the main clinical reason for interventional catheterizations in the early postoperative period as it was cyanosis in the late follow-up. Hemoptysis was the most frequent cause for cardiac recatheterizations. A broad spectrum of percutaneous procedures was performed. Mortality was mainly related to unstable hemodynamic status but not to the interventional procedure in itself. Interventional percutaneous procedures are mandatory

P1705 - CARDIAC MECHANICS IN CHILDREN POST PERCUTANEOUS TRANSCATHETER CLOSURE OF PERIMEMBRANOUS VENTRICULAR SEPTAL DEFECT RIGHT AND LEFT VENTRICULAR PERFORMANCE ASSESSED BY ECHOCARDIOGRAPHY

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Background: Percutaneous transcatheter closure of perimembranous ventricular septal defect (pmVSD) with occluder has been

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most widely used in China. However, whether impaired ventricular performance occurs in pediatric patients post transcatheter closure of pmVSD, is not known. Two-dimensional speckle-tracking echocardiography (2D-STE) is increasingly used to evaluate ventricular performance in children. In this study, we aimed to analyze left ventricle (LV), right ventricle (RV) performance post percutaneous transcatheter closure of pmVSD.
Methods: 40 pediatric patients post percutaneous pmVSD closure and 40 healthy children were recruited. All subjects were studied with conventional and tissue Doppler echocardiography. Strain and strain rate of LV and RV were assessed by 2D-STE.
Results: 40 pmVSD patients (6.0 ± 1.2 years) and 40 healthy controls (5.8 ± 1.1 years) were studied. Mean diameter of pmVSD was 3.82 ± 0.59 mm, mean diameter of pmVSD occluder was 6.3 ± 1.0 mm, and mean time after percutaneous pmVSD closure was 3.22 ± 0.78 years. No significant differences were observed in LV eject fraction, RV Tei index and transatrioventricular velocity between pmVSD closure and control. More tricuspid regurgitation was observed in pmVSD closure subjects by measuring the ratio of tricuspid regurgitation jet area and right atrial area (TRJA/RAA) ($p = 0.009$). Interventricular septal tissue Doppler image showed less early diastolic ($p = 0.01$), more late diastolic ($p = 0.04$) velocity and less e/a ratio ($p = 0.005$) in pmVSD closure subjects. No significant difference in global longitudinal and circumferential strain and strain rate between pmVSD closure and control. For RV deformation, pmVSD closure subjects tended to be lower global longitudinal strain($p = 0.06$). For pmVSD closure cohort, the diameter of pmVSD occluder correlated negatively with LV longitudinal strain rate ($r = -0.324$, $p = 0.044$) and circumferential strain ($r = -0.354$, $p = 0.027$). Furthermore, TRJA/RAA correlated positively with diameter of pmVSD ($r = 0.727$, $p < 0.001$) and occluder ($r = 0.777$, $p < 0.001$).
Conclusion: 2D-STE appears that percutaneous closure of pmVSD is safe and effective in selected pediatric patients.

P1716 - EARLY POST OPERATIVE INTERVENTIONAL CATHETERIZATION OF CONGENITAL HEART DEFECTS SHORT TERM RESULTS

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Introduction: Interventional cardiac catheterization is an alternative to surgery in the management of residual postoperative defects. There are very few studies published in the literature that evaluate its efficacy, safety and describe its complications.
Materials and Methods: We performed a retrospective study over a period of 5 years in patients under 18 years of age, who underwent catheterization in the first 30 days after the congenital heart surgery. Data on demographic, clinical, diagnostic, surgical, hemodynamic, required support, complications and 30-day survival were collected to determine the types of procedures, outcomes, and complications.
Results: 42 catheterizations were performed in 34 patients, most of them male (70.6%), with a median age of 183 days and weight of 6 Kgrs. The main surgical procedures were the truncal corrections followed by Norwood surgeries. A median of 6 days elapsed between surgery and catheterization, with hypoxemia, dependence on ventilatory assistance and suspicion of obstructions in the operated areas being the main indications of catheterization. 62% were interventional catheters, with angioplasty being the most frequent, with balloon (54%) and stent (27%). The most affected sites were the pulmonary arteries (57%), followed by the aortic arch (11%) and the cavus pulmonary junction (9%). Complications were recorded in 7% (5 patients): Arrhythmias in 3 patients, need for ECMO in 1 case and 1 with cardiac arrest. There were no deaths related to the procedure, the survival at 30 days was 84%. The need for inotropes and mechanical ventilation was reduced in more than 50% of the cases.
Conclusions: Early diagnostic and therapeutic post-surgical catheterization can be performed with low mortality and morbidity and with adequate efficacy.

P1720 - LONG TERM PROGNOSIS OF TRIPLE BALLOON ALLOPLASTIC IN PATIENTS WITH PULMONARY VALVE STENOSIS

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Percutaneous pulmonary valved stents implantation for patients with right ventricle outflow tract stenosis is now considered feasible and safe, however, there are some limitations. Triple balloon valvuloplasy for these patients has been reported its efficacy and safety, however, the long-term prognosis of patients is still unknown, especially whether pulmonary valve regurgitation is getting worse or not. Here, we analyzed the long-term prognosis of patients receiving triple balloon valvuloplasty. Five patients (median age 31years, 4 females, 1 male, tetralogy of Fallot after surgical repair 2, ventricular septal defect after surgical repair 1, native pulmonary valve stenosis 2) underwent the procedure safely and successfully. The median diameter of pulmonary valve annulus was 23.0 mm, and balloon diameters ranged from 12 to 15 mm. The median peak right ventricular-to-pulmonary artery systolic pressure gradient was 40.0 mmHg before intervention and was reduced to 10 mmHg after that. No major complication was occurred. The median follow-up period was 73.5 months. There were no patients who got worse in pulmonary valve regurgitation after the procedure. Four patients were no need for additional operation during observation period. One patient who had residual shunt after intracardiac repair of ventricular septal defect was needed surgical repair because the left-to-right shunt was increased. Triple balloon valvuloplasty is effective and safety procedure, and can be useful as a palliative therapy because of small risk of the deterioration in pulmonary valve regurgitation.

P1732 - BALLOON PULMONARY VALVULOPLASTY SINGLE CENTER EXPERIENCE AND LONG TERM OUTCOME

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Background: Balloon pulmonary valvuloplasty (BPV) is the treatment of choice for pulmonary valve stenosis. Besides its effectiveness, the long-term outcome have to be better understand.
Objective: To describe a single-center experience and long-term outcome with BPV.
Methods: A retrospective study between 2001 and 2016. The catheterization laboratory records from all patients who underwent BPV were reviewed and compared with the recent echocardiography findings.

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Results: From November/2001 to November/ 2016, eighty-five patients (Pts) underwent BPV. Thirty-four patients were males (40%), the median age was 4 years (2 days ~ 61 years). Forty-eight (56%) patients were younger than 1 year and within this group, twenty-two (46%) were newborns. The mean peak systolic pressure gradient across the pulmonary valve in the whole group decreased from 65.4 to 16.8 mmHg (136 mmHg ~ 35 mmHg) and from 56 to 12.0 mmHg in the group of patients <1 yr. BVP was successful in 90% of the cases with a global mortality of 1.9%. Mild complications occurred in 9 patients (11%), all of them were in the group <1 yr: arrhythmia (4), infundibular spasm (3), cardiac tamponade (1), dissection of IVC (1). Thirty –three patients (65%) with >5 year follow-up had an echocardiogram performed, twenty-five (75%) of them presented pulmonary regurgitation (PR). Twelve (36%) with moderate/important PR, thirteen (40%) with trivial and eight (24%) without PR. The balloon/annulus ratio >1.4 was seen in 75% in of Pt with moderate/important PR, 23% with trivial and 30% without PR.
Conclusion: BPV is a safe and effective procedure for pulmonary valve stenosis. The balloon/annulus ratio >1.4 seemed to be a prediction of PR in this study. Further studies with MRI may be useful for better assessment of the right ventricular function on the follow-up of these patients.

P1747 - TRUE RISK FACTORS FOR LOSS OF ARTERIAL PEDAL PULSE FOLLOWING CARDIAC CATHETERIZATIONS IN INFANTS

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Background: The objective of this study was to compare the prevalence of acute loss of pedal pulse (LOP) in infants (≤1 year) who had ultrasound guided femoral arterial access (UGFAA) during cardiac catheterizations between three time periods: Period-1 (Jan2003–Dec2011), Period-2 (Jan2012–Dec2014) and Period-3 (Jan2015–Dec 2016). A prospective study was previously conducted during Period-2. The diameter of the accessed femoral artery (FA) <3mm was found to be the only independent predictor for LOP. Since then, we attempted to avoid FA cannulation if the vessel size was <3mm and use smaller catheters if FA access was necessary during Period-3.
Methods: A retrospective study was performed including 808 cardiac catheterizations in infants, 516 in Period-1, 166 in Period-2 and 124 in Period-3. LOP was defined as a loss of palpable pedal pulse at the end of the procedure. The FA size was measured prior to USGFAA and a ratio of the outer diameter of the arterial sheath to the luminal diameter of the cannulated artery (OD/AD ratio) was calculated during Periods-2&3.
Results: There was a significant decrease in the FA access rates during Period-3 (64.5% vs. 93.8% during Period-1 and 90.4% during Period-2; $P < 0.001$). This coincided with a decrease in the prevalence of LOP during Period-3 to 3.5% compared to 13.4% during Period-1 and 15.7% during Period-2 ($P < 0.001$). By multivariate analysis, smaller FA size and a larger OD/AD ratio were the only significant independent predictors for LOP (OR:6.48, 95% CI: 2.31–11.42, $P < 0.001$ and OR:4.16, 95% CI: 1.79–8.65, $P < 0.01$, respectively).
Conclusions: Limiting access of FA <3mm during cardiac catheterizations was associated with a significant decrease in the prevalence of LOP in infants. This study validates the findings of our previous study that smaller FA size and a larger OD/AD ratio are the only significant predictors for LOP in infants.

P1749 - CHANGE IN LIMB TISSUE PERFUSION DURING CARDIAC CATHETERIZATION UTILIZING THE FEMORAL ARTERY IN CHILDREN

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Background: Arterial access during cardiac catheterization in children can effect distal limb tissue perfusion (Fdl); the risk factors for which are unknown. The objectives of this study were to measure changes in Fdl and identify risk factors for decreased Fdl in children during cardiac catheterization.
Methods: This is a prospective, single center, pilot study of distal limb perfusion during pediatric cardiac catheterizations from August-November 2016. Laser Doppler flowmetry was used to continuously measure Fdl using real-time, tissue RBC Fluxes. The percentage difference in the pre-to-immediate post-procedure Fdl (ΔP-Fdl) and the pre-to-lowest Fdl during the procedure (ΔL-Fdl) were calculated. A 6-hour post-procedure Fdl (6h-Fdl) was also measured. The ratio of the outer diameter of the arterial sheath (OD) to the luminal diameter of the cannulated artery (AD) OD/AD ratio was calculated. Risk factors for the greatest change in Fdl during a procedure (highest ΔL-Fdl) were determined.
Results: 50 children (median age = 15 months, median weight = 10 kg) were included in the study. The ΔP-Fdl ranged between 0% and 85% (median 22.4%), and the ΔL-Fdl ranged between 17.5% and 96.8% (median 36.2%). An OD/AD > 50% was the only significant predictor for greatest change in Fdl during a procedure (OR:4.6, 95% CI: 2.7–11.4, $P < 0.001$). The luminal diameter of the cannulated artery, procedure length and pre-post procedure change in AD were not found to be significant predictors. Pulse loss was not encountered in any patient. The 6h-Fdl was 0-21% (median = 7%) lower than the pre-procedure Fdl ($P = 0.34$).
Conclusions: This pilot study is the first to determine real-time changes in distal limb tissue perfusion in children during cardiac catheterization using arterial access; the clinical implications of which are still unknown. The knowledge that an OD/AD ratio >50% could significantly affect Fdl may be useful for practitioners in preventing any long term sequelae from these procedures.

P1752 - TRANSCATHETER IMPLANTATION OF SAPIEN XT VALVE WITHOUT PRE STENTING IN RIGHT VENTRICULAR OUTFLOW TRACT HOMOGRAFT FOR SEVERE PULMONARY REGURGITATION FOLLOWING TETRALOGY OF FALLOT REPAIR

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Background: Surgical reconstruction of right ventricle outflow tract (RVOT) is often performed in patients undergoing repair of tetralogy of Fallot (TOF). Pulmonary homograft valve deterioration is expected over time. Previously, correction of pulmonary insufficiency or stenosis required repeat cardiac surgery. Percutaneous pulmonary valve implantation (PPVI) has emerged as a viable alternative in patients with RVOT dysfunction. The Sapien XT system has recently been approved for PPVI with conduit prestenting necessary.

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Case report: A 24 year old female with TOF and absent pulmonary valve syndrome underwent complete surgical repair in 1998; Dacron patch closure of the VSD, 21 mm pulmonary homograft reconstruction of the RVOT. She had chronic, hemodynamically significant, pulmonary regurgitation with RV volume overload and required right pulmonary artery (RPA) stent angioplasty in 2013. Her clinical status worsened with increasing fatigue, disproportionate increase in RV size, and development of arrhythmia; but she declined surgery. Her minimum conduit diameter is 20.3 mm, annulus 23.3 mm and aneurysmal RVOT proximal to homograft is 34.8 mm. Balloon stretch diameter of the homograft potential landing zone is 22 × 23 mm. However, anatomy was unsuitable for pre-stenting due to 9-12 mm distance to the mouth of the RPA stent, thus precluding use of a Melody valve. Instead, she had transcatheter Sapien XT 26 mm valve implantation, delivered via an 18 Fr delivery system, without pre-stenting to avoid compromising flow into the RPA. The Sapien valve remained in stable position with no valvular or paravalvular leaks identified and trivial obstructive gradient.
Discussion: The Sapien XT transcatheter valve serves as an excellent adjunct to therapy in congenital pulmonary valvular disease and high success rates have been demonstrated. We report a case of Sapien XT PPVI implantation without pre-stenting with successful outcome as demonstrated by resolution in pulmonary regurgitation and no compromise to flow into the RPA.

P1778 - IN VIVO AND IN VITRO TESTING FOR HAND MADE TRILEAFLETS PERICARDIAL PATCH VALVED CONDUITS IN PULMONARY VALVE RECONSTRUCTION

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Background: The hand-made trileaflets pericardial patch valved-conduits might be used as the pulmonary valve replacement substitute materials. The aim of this study was to investigate the valve function of the hand-made trileaflets pericardial patch valved-conduits by mock system (in vitro) and their short-term conditions in piglet animal model (in vivo) to justify the applicability of clinical purpose.
Methods: The competency of the hand-made trileaflets pericardial patch valved-conduits were tested on the pulmonary circulation mock system. The valved-conduit assembled into a delivery system to deploy into the pig animal destroyed pulmonary valve model via femoral vein under general anesthesia. In vivo functional and morphology analysis were tested during a 4-week period.
Results: On mock circultatory system, the hand-made trileaflets pericardial patch valved-conduit, although not as perfect as the silicon trileaflets valve, can effectively rise the mean pulmonary pressure from 10.2 to 14.4 mmHg with compared to the defected silicon valve one. The regurgitation fraction (RF) value of hand-made trileaflets pericardial patch valved-conduit group is 15.9-18.1% significantly improved compare with the defected valve group (RF = 73.5-85.7%). The in-vivo pulmonary condition experiments showed their good valve position conditions and the valve and leaflets characteristics and function. The histological analysis of the valved-conduits adjacent pulmonary area also revealed no significant signs of inflammation reaction.
Conclusion: The hand-made trileaflets pericardial patch valved-conduits for pulmonary valve reconstruction on both mock and animal experiments showed with acceptable performance and outcome. It might implicate that this hand-made trileaflets pericardial patch valved-conduits can be used in further clinical purpose. However, the long-term animal results still need time to justify.

P1798 - IS ADO II IDEAL FOR CLOSING CONGENITAL GERBODE DEFECTS

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Background: Congenital left ventricle to right atrial communications (Gerbode defects) are extremely rare (0.08%) type of ventricular septal defects. They were traditionally closed by surgery in the past. There are few case reports and small series of acquired and congenital Gerbode defects, closed with various types of devices. We report a series of only congenital Gerbode Defects closed by Amplatzer duct occluder II (ADO II). Aim of our study is to assess the feasibility, efficacy, and complications of transcatheter closure of congenital Gerbode defects with ADO II.
Material and Methods: 13 consecutive cases of Gerbode defects, age ranging from 10 months to 16 years (mean 6.7 years), weight ranging -6.5 kg to 34 kg (mean 19.3 kg), were diagnosed on transthoracic echocardiography and left ventricular angiogram done in AP, RAO view with 10 0 cranial angulation, were closed with ADOII by retrograde approach The fluoroscopic time was 6.2 +1.4 min.
Results: Transcatheter closure of Gerbode defects,(both type A and B) with ADO II was successful in all 13 cases (100%). Only one patient had transient complete heart block, made a complete recovery with temporary pacing for 48 hours and steroids for five days. No aortic or tricuspid regurgitation or residual shunt occurred.
Discussion: Soft low profile, easily trackable ADO II appears to be ideal for closure of Gerbode defects, as the central cylinder fits in the defect and the soft retention discs on either side, without polyester material, do not impinge on either aortic, mitral, or tricuspid valve.
Conclusions: Transcatheter closure of congenital Gerbode defects with ADO II is safe, effective, and an attractive alternative to surgical closure. ADO II appears to be tailor made for Gerbode defects, as the success rate is 100%

P1799 - TRANSCATHETER INTERVENTIONS IN APICAL NON COMPACTION TO REDUCE THE PUMP FAILURE

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Background: Apical non-compaction (ANC) of both ventricles and septum is not reported much in literature. For the first time, we report interventions for associated lesions in ANC.
Aim: To know feasibility of transcatheter interventions for associated lesions in ANC to reduce the pump failure.
Material and Results: Out of 100 consecutive patients of ANC 36 cases underwent various transcatheter interventions, formed the material. Age ranged 3days to 17 years (mean 8 years). Device closure was done for 2 PDA, 16 VSDs, 1 ASD, one each of ALVT, ARVT, ARAT, 5 ABV, 2 PBV, 1 aortoplasty, 2 PTMC, I for

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CTD, 3 pericardiocentesis. 5 patients underwent two procedures in the same sitting eg ABV and PBV, ABV and PTMC, ABV and VSD device closure, ASD and VSD device closure and PDA and VSD device closure. 3 cases of VSD were post- operative residual shunts and one was closed with multiple devices. One 8 months infant had apical VSD closed with ADO II. 2 year old child underwent hybrid surgery for closure of VSD with 14 mm device. Mid muscular VSD was closed with ADO II in mirror image dextrocardia.
Discussion: Interventions in ANC are challenging in presence of LV/RV dysfunction with or without thrombosis. Positioning device in apical VSD in ANC cases is very challenging as the device gets caught in trabeculae in RV and if more tug is given the device slips through spongy myocardium. The results of interventions are very gratifying as pump failure due to pressure or volume overload caused by associated lesions improves significantly.
Conclusion: Transcatheter interventions though challenging are feasible safe effective and are lifesaving. Transcatheter interventions certainly reduce the morbidity and mortality in ANC patients who are at high risk for surgery or redo surgery.

P1822 - PERCUTANEOUS CLOSURE OF PDA IN BABIES BELOW 7KG WHAT WE HAVE LEARNED IN THE LAST YEARS

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Background: Percutaneous closure of patent ductus arteriosus (PDA) is well established. However, most of low weight infants are still sent to surgery. Our aim is to show our experience with percutaneous treatment of children below 7 kg in the last 5 years.
Material and Methods: We reviewed all medical records of patients who underwent percutaneous occlusion of PDA from January 2011 to August 2016. We analyzed characteristics of the patient (sex, age, weight and clinical status), the ductus (Krichenko classification and size at the pulmonary end) and the procedure (type and size of the device, hospitalization time, complications).
Results: Of 252 patients referred for PDA occlusion, 15 (5,9%) had less than 7 kg. Eight were female and 7 male. Ages ranged from 2 months to 1 year of age and weights from 5 to 7 kg. Twelve patients (80%) had signs and symptoms of congestive heart failure (CHF). Three patients had Down syndrome and 4 had other cardiac malformations. Extracardiac malformations were found in 2 patients. Most PDAs were Krichenko type A, although 2 were type C. The pulmonary end of the PDA ranged from 2 to 6mm (median 4mm). We were able to implant devices in 14 patients. Of these, 6 received AMPLATZER devices of various types, 7 CERA devices and 1 NIT OCCLUD PDA-R device (as displayed in the graph). Only one patient had residual shunt after 6 months of follow-up. There was no obstruction of the descending aorta or of the pulmonary branches, no vascular complications and no death related to the procedure.
Conclusion: Infants weighing less than 7 kg with PDA often have CHF and other comorbities, therefore closure is urgent and a less invasive treatment is more suitable. We concluded that the procedure is safe, especially if we can choose the most appropriate device for each case.

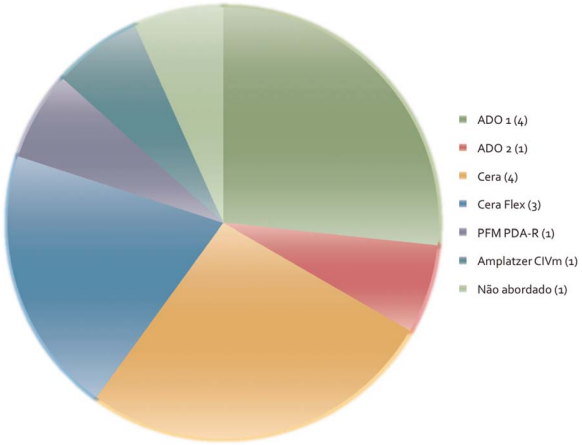

Figure.

P1829 - ATRIAL SEPTAL DEFECT (ASD) IN SMALL CHILDREN DO WE NEED TO WAIT TO CLOSE THEM

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Background: ASDs often cause right ventricular overload, which progressively gets worse and eventually progresses to pulmonary hypertension. As soon as a cardiac malformation is diagnosed, the family is anxious for it to be resolved. Therefore, we believe that ASDs should be closed as soon as possible. Our aim was to demonstrate that small patients (below 18 kg), percutaneous occlusion of ASD is feasible.
Material and Methods: We reviewed clinical records of all ASDs closed percutaneously from January 2011 to August 2016. We analyzed sex, weight, age, clinical and echocardiographic features, size and type of device and complications related to the procedure.
Results: In this 5-year period, we closed percutaneously 158 ASDs. Of these, 23 (14,4%) were in patients with less than 18 kg; 15 female and 8 male. Age ranged from 3 to 7 years and weight from 13 to 18 kg. All of them were referred to treatment because of right ventricular overload. There were 3 cases of aneurysmal septum with multiple defects; 2 of our residual shunts were in this group. The devices we used were AMPLATZER ASD (4), OCCLUTECH FIGULLA (12), NIT OCCLUD ASD-R (5) and CARDIA ATRIASEPT (2). We performed balloon measurement in all cases; stretched diameter ranged from 10 to 24mm. The size of the device chose varied from 4% to 36% bigger than the measured diameter. We had no vascular complication and no death. We were aided by transesophageal echo (TEE) in all cases, and there was no complication related to the probe. Until the present date, there was no erosion and no arrythmia in this group.
Conclusion: Moderate and large ASDs with right ventricular overload can be approached early, with excelent results and no complications in short and medium term follow-up.

P1842 - DUCTAL STENTING AS AN ALTERNATIVE TO THE BLALOCK TAUSSIG SURGERY

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Background: The Blalock-Taussig (BT) shunt is a life-saving surgery, but it still presents significant morbidity and mortality. In this scenario, stenting the ductus arteriosus (DA) appears as a less invasive alternative. Our goal is to describe the results of stent implant in place of BT as a palliation in children with cyanotic malformations.
Material and Methods: A review of the charts of all intended stent implantation from 2008 to 2016 was carried out. We analyzed the following characteristics: sex, age, weight, diagnosis, anatomical features of the ductus, causes for failure and complications.
Results: Twenty-nine cases of attempted stent implantation were surveyed, with a success rate of 82,8% (5 failures). Seventeen patients were male and 12 female. Ages ranged from 5 days to 16 months and weights from 2.4 kg to 10.4 kg. The echocardiographic diagnosis were: pulmonary atresia and ventricular septal defect (13 patients), pulmonary atresia with intact ventricular septum (9), severe pulmonary stenosis (3), tricuspid atresia (1), double outlet right ventricle with pulmonary stenosis (1), pulmonary atresia with single ventricle and right isomerism (1) and tetralogy of Fallot (1). As to the characteristic of the ductus, 6 were horizontal and straight, 2 horizontal ant tortuous, 10 vertical and straight and 11 vertical and tortuous. There were 2 cases of pericardial effusion, one of them in the success and the other in the failure group. Of the cases with successfully implanted stents, there were 3 deaths, one due to cardiogenic shock and two due to sepsis. Among the 5 cases of failed implantation, there was 1 death in the same day of the procedure and the other 4 patients were referred to BT surgery. The failure cases were due to ductus tortuosity or stenosis.
Conclusion: The results show that stent implantation is a good alternative to BT surgery in cyanotic patients who present a favorable ductus morphology.

P1846 - OCCLUSION OF WINDOW LIKE DUCTUS WITH ATRIAL SEPTAL DEVICE

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Background: Percutaneous occlusion of Krichencko type B ductus arteriosus (DA), also called window-like ductus, is a challenge to the interventional cardiology. Devices designed for ductus closure are unsuitable for this morphology, with a high risk of displacement or protrusion to the aorta or the pulmonary artery. We report a case of percutaneous closure of a type B ductus successfully performed with an atrial septal defect (ASD) device.
Case Report: Our patient was a 4 month-old boy, weighing 4.8 kg with Down syndrome and a history of fatigue during breastfeeding. Physical examination showed tachypnea, tachycardia, oxygen saturation of 96%, and a loud systolic murmur. Electrocardiogram showed signs of right atrial and right ventricular enlargement. The first echocardiogram showed a small ASD, a moderate DA and a pulmonary artery pressure (PAP) estimated by the tricuspid regurgitation of 50 mmHg. He was initially treated with diuretics. Nine months later, his saturation were lower (85%) and the PAP was 80 mmHg. In this stage, he was given SILDENAFIL and referred to intervention. During the procedure, we were able to document a fall in the PAP after temporary occlusion of the DA with a balloon. We then proceeded to successfully implant the OCCLUTECH FIGULLA ASD FLEX II size 6. The patient was discharged the following day and has been asymptomatic.
Coments: Success of the percutaneous closure of the window-like DA brings the possibility of correction of this defect using a method less invasive, with less complications and lower hospitalization time.

P1850 - TRANSCATHETER TREATMENT OF COARCTATION OF AORTA AND DUALLY CONNECTED ANOMALOUS VERTICAL PULMONARY VEIN

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Aga Khan University Hospital, Pediatrics and Child Health, Karachi-Pakistan[1]; Aga Khan University Hospital, Cardiac Surgery, Karachi-Pakistan[2]

Introduction: Partial anomalous pulmonary venous connection is failure of one or more pulmonary veins to connect with left atrium during fetal development. It is frequently associated with atrial septal defects, and rarely with other congenital cardiac abnormalities. We describe transcatheter treatment of aortic coarctation and occlusion of partial anomalous pulmonary venous connection in the same patient.
Case Report: Thirteen year old girl was diagnosed to have coarctation of aorta an anomalous left upper pulmonary vein to innominate vein. She was planned to have the coarctation treated with covered stent and the anomalous vein treated surgically at a later date. Cardiac catheterization and angiography showed juxtaductal coarctation, narrowest segment measured 3.8 mm with a pressure gradient of 40 mmHg across the coarctation. Oxygen saturations were 99% in the pulmonary veins, 93% in the innominate vein and pulmonary artery, and 82% in the proximal superior vena cava. Qp /Qs was 4.5. Left pulmonary angiogram showed normal pulmonary venous connection to left atrium with a large vertical venous channel connecting the confluence of left pulmonary veins to innominate vein. Balloon occlusion of the vertical vein did not interfere with left pulmonary venous drainage. Coarctation of aorta was treated with NuMED covered CP stent, 39mm X 8 zig mounted over a 12mm X 4 cm Z Med balloon reducing the gradient to 4 mmHg. Vertical vein measured 13 mm in diameter and was closed with 20 mm Amplatzer vascular plug II, released after confirming unobstructed pulmonary venous return.
Discussion: Historically, multiple cardiac pathologies, such as in the present case, would be treated with surgery. Improved technics have made it possible to address multiple defects by percutaneously at the same time. In our patient we stented the coarctation first and then addressed the vertical vein, which was possible due to dual pulmonary venous connection.

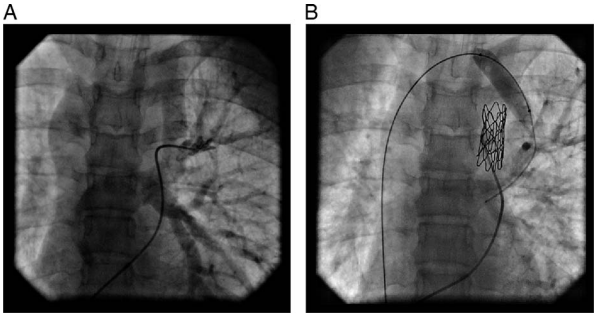

Figure 1.
Anomalous vertical vein with dual pulmonary venous connection 9A0 and balloon occlusion (B) with coarctation stent in place.

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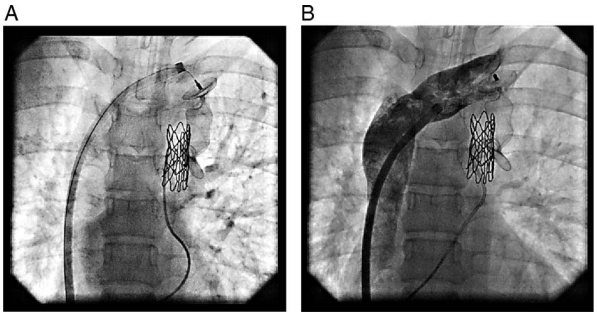

Figure 2.
Deployment of Amplatzer vascular plug II (A) and release(B).

P1871 - PROLONGED CARDIAC CATHETERIZATION THE INFLUENCING FACTORS AND SEQUENTIAL TIME ANALYSIS

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Background: Cardiac catheterization has developed worldwide. Apart from success rate, the procedural time is an important parameter for efficacy. Shorter duration could reduce costs and risk of complications. This study aims to identify the influencing factors and sequential times for prolonged cardiac catheterization.
Material and Methods: The retrospective study conducted from November 2009 to November 2016 at Suratthani Hospital, Thailand. Pediatric and adult patients who underwent cardiac catheterization under general anesthesia were included in the study. Demographic data, types of procedures, and the procedural time for all processes were recorded. Prolonged cardiac catheterization was defined as cases in which procedural time was greater than the mean time. Identification of the influencing factors and comparisons of the times of all processes between groups were assessed using t-test and Chi-square test. A statistically significant level was defined by p-value <0.05.
Results: Of 140 patients, 68 had prolonged cardiac catheterization (the mean time was 101.85 ± 35.54 minutes). Of the four comparative factors: age ≤ 1 year, body weight ≤ 10 kilogram, cyanotic heart diseases and interventional cardiac catheterization, age ≤ 1 year is an influencing factor on prolonged cardiac catheterization ($p = 0.008$). A sequential time analysis revealed a higher preparation time by nurses and anesthesiologists, catheterization time, and post-procedural time in the prolonged cardiac catheterization group compared with the other group. These parameters were also significantly longer in patients aged ≤ 1 year.
Conclusions: Age ≤ 1 year is an influencing factor on prolonged cardiac catheterization which was affected by multiple processes. Although low age is difficult to avoid in patients with absolute indications for cardiac catheterization, the procedure should be delayed in elective cases. Recognition of the causes of prolonged catheterization in each processes and implementation of more effective methods could reduce the procedural time and increase the efficacy of cardiac catheterization.

P1904 - TRANSCATHETER STENTING AND CLOSING A DUCTUS IN A NEWBORN WITH BILATERAL DUCTUS

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National Cheng Kung University Hospital, Pediatrics, Tainan-Taiwan

Background: With the advance of transcatheter technique, transcatheter closure or stenting of a ductus is now regarded a safe and feasible procedure in selected infants.
Materials and Methods: We report a newborn patient (birth body weight: 2280 g) who presented with dyspnea and cyanosis 6 hours after birth. Differential cyanosis (SpO2 of right arm: 98%; SpO2 of right leg: 83%). Chest plain film revealed cardiothoracic ratio: 56% and uneven pulmonary vascular markings between bilateral lung. Echocardiogram showed a perimembraneous ventricular septal defect, main pulmonary trunk giving rise to right pulmonary artery and large right ductus arteriosus, left ductus arteriosus which arises from aortic arch close to the origin of left brachiocephalic artery connecting to left pulmonary artery, interruption of right and left pulmonary artery, right arch, and hypoplastic aortic ach. Majority of flow from pulmonary trunk goes to descending aorta through right ductus arteriosus. Considering high risk of total repair, we performed palliation procedure for this patient.
Results: Our strategy is balloon angioplasty for hypoplastic arch if significant pressure difference is present between ascending and descending aorta, transcatheter closure of right ductus, and placement a stent in left ductus. During cardiac catheterization, there was no pressure gradient between ascending and descending aorta. Amplatzer® vascular plug 10mm in diameter was used to occlude right ductus. We put an OMEGA™ stent, 4.0mm*8.0mm, in left ductus to promote left pulmonary artery growth. This patient was discharged soon without complications.
Conclusions: With the advance of transcatheter technique, closure or maintenance of patency of ductus can be both achieved without surgery. We report our experience in both closing and stenting a ductus through cardiac catheterization in a newborn with complex congenital heart disease.

P1915 - STEPWISE APPROACH TO CONDUIT PUNCTURE FOR ELECTROPHYSIOLOGICAL PROCEDURES IN PATIENTS WITH FONTAN CIRCULATION

Jae-Sun Uhm[1], Nam Kyun Kim[2], Jung Ok Kim[2], Jae Young Choi[2], Moon-Hyoung Lee[1], Jo Won Jung[2], Hui-Nam Pak[1], Boyoung Joung[1], Tae-Hoon Kim[1], Sang-Yun Lee[3], Mi Kyoung Song[4]
Severance Cardiovascular Hospital, Yonsei University College of Medicine, Cardiology, Seoul-Korea, South[1]; Severance Cardiovascular Hospital, Yonsei University College of Medicine, Pediatric Cardiology, Seoul-Korea, South[2]; Sejong General Hospital, Bucheon, Pediatric Cardiology, Gyeonggi-Do-Korea, South[3]; Seoul National University Children's Hospital, Pediatric Cardiology, Seoul-Korea, South[4]

Background: Electrophysiological procedures are challenging in patients with Fontan conduit,the conduit should be punctured for electrophysiological procedure.
Methods: In our study, 12 patients with lateral tunnel or extracardiac conduit Fontan circulation [median age (interquartile range), 21.5 (16.0–25.3) years; 6 men] who had undergone electrophysiological procedures were enrolled. We performed stepwise approach to Fontan conduit puncture; 1st step, Brockenbrough needle; 2nd step, Brockenbrough needle + snare; 3rd step, Brockenbrough needle (XS series) ± snare; 4th step, radiofrequency transseptal needle ± snare; 5th step, wiring through the puncture; 6th step, conduit dilation with non-compliant balloon; 7th step, cutting balloon; 8th step, Inoue dilator.
Results: In 2, 1, and 2 patients with lateral tunnel made of the pericardium or right atrial wall, conduit puncture was performed by step 1, 2, and 4, respectively. In 2, 1, and 1 patient with the Goretex lateral tunnel or extracardiac conduit, conduit puncture was performed by step 6, 7, and 8, respectively. Puncture time was

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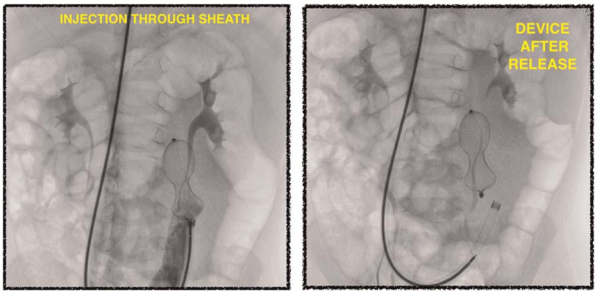
significantly longer in patients with Goretex conduit than in patients with pericardial conduit [91.0 (59.8–130.5) and 11.5 (10.0–14.8) minutes, respectively; $p < 0.001$].
Conclusions: Stepwise approach to conduit puncture is feasible and safe in patients with lateral tunnel and extracardiac conduit Fontan circulation. Goretex conduit puncture is more difficult than pericardial conduit puncture.

P1924 - ABSENT PORTAL VEIN AND BLEEDING HAEMORRHOIDS IN A TWO YEAR OLD CHILD POSTED FOR LIVER TRANSPLANTATION

Sudeep Verma[1], Gouthami V[2], Raja Mannar[3], Sethu Babu[4], Nagarajan R[5], Sachin Daga[6]
Kims Hospital, Pediatric Cardiac Sciences, Hyderabad-India[1]; Kims Hospital, Peddiatric Cardiac Sceinces, Secunderabad-India[2]; Kims Hospital, Intervention Radiology, Secunderabad-India[3]; Kims Hospital, Department of Gastroenterology, Secunderabad-India[4]; Kims Hospital, Pediatric Cardiac Anesthesia, Secunderabad-India[5]; Kims Hospital, Departement of Surgical Gastrocenterology, Secunderabad-India[6]

Background: Haemorrhoids is rare in paediatric age. Presenting a rare case of bleeding per rectum due to haemorrhoids secondary to portal hypertension and congenital absence of portal vein (Abernathy malformation I) referred for liver transplantation. Balloon occlusion of the aberrant vessel demonstrated presence of portal radicals, followed by successful closure of the aberrant channel. Follow up showed good growth of the portal radical, otherwise reported as absent during initial assessment.
Description: 2 years old, weighing 10.5 kg presented with complaints of recurrent bleeding per rectum. Examination showed severe pallor and splenomegaly. Suspected to have portal hypertension hence CT scan and MRI was done that showed congenital absence of portal vein with abnormal drainage of portal confluence to right iliac vein (Abernathy type I - Morgan classification and Kobayashi classification type C). Haemoglobin was 7 gm% and LFT were normal. Sigmoidoscopy revealed large venous channel into rectum with multiple internal haemorrhoids. Balloon occlusion test of the draining channel using 16×40 TYSHAK BALLOON demonstrated presence of portal radicals to the liver. Portal pressure measured 19 mm Hg after test occlusion. Device closure of the aberrant channel was done using 24/26 duct Occluder from right jugular vein route. Ammonia level dropped from 95 to 20 after the procedure. Eight months follow up showed decreased rectal bleeding and stable haemoglobin level of more than 11 gm%. Follow up angiogram showed good growth of portal radical to liver with normal LFT.
Conclusion: Bleeding haemorrhoids is a rare presentation of portosystemic shunt. Diagnosis of Absent portal vein should always be consider only after balloon occlusion test. Surgical ligation or transcatheter closure of aberrant vessel is feasible option if portal pressure remained less than 25-30 mm Hg. Long term follow up is needed to assess portal venous pressure, growth of intrahepatic portal radicals.



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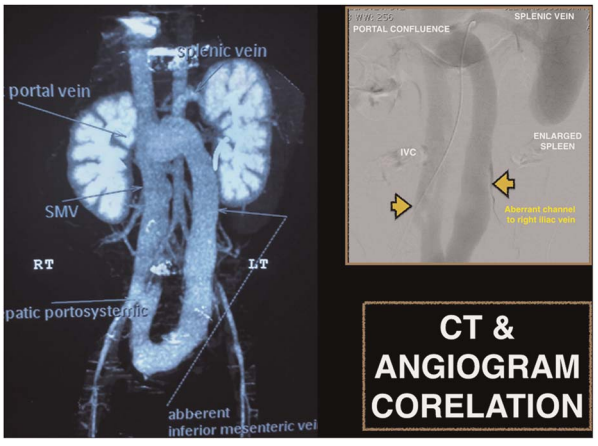

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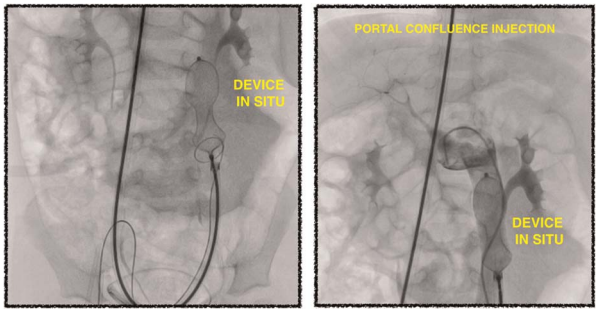

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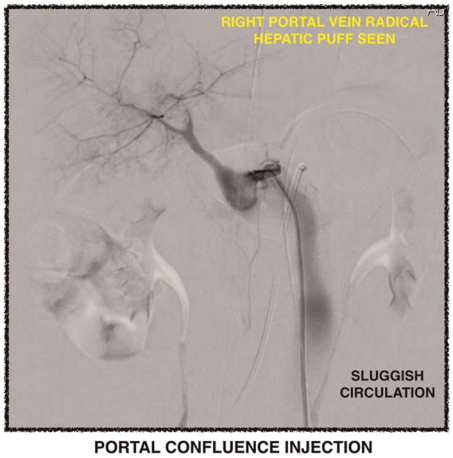

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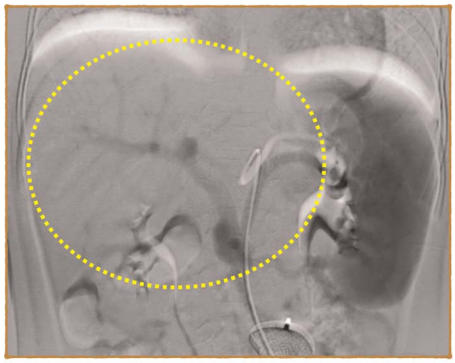

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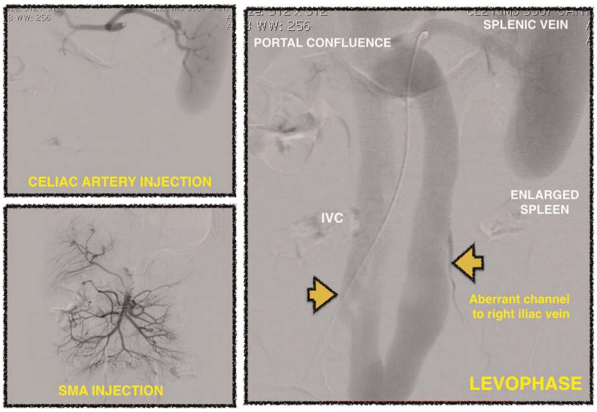
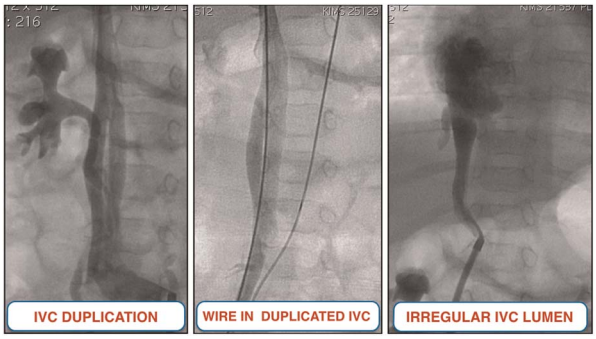


Figure 6.

P1925 - INFRARENAL IVC DUPLICATION AND DECOMPRESSING SPINAL VEINS AS A CAUSE FOR SEVERE IVC SPASM IN A EUVOLEMC PATIENT DURING PDA DEVICE CLOSURE

Sudeep Verma[1], *Gouthami V*[1], *Nagarajan R*[2], *Anil Dharmapuram*[1], *I.M Rao*[1]
Kims Hospital, Pediatric Cardiac Sciences, Hyderabad-India[1]; *Kims Hospital, Pediatric Cardiac Anesthetist, Secunderabad-India*[2]

Background: Spasm of the vessel is a known entity during manipulation of hardwares during catheterization. We are describing a case where severe IVC spasm happened during PDA device closure in a euvolemic patient precluding catheter to advance into RA. Evaluation revealed infrarenal duplication of IVC and filling of left paraspinal venous plexus causing decreased effective lumen of the IVC. This case highlights the importance of suspecting IVC abnormality in case of unusual spasm of the major vessels.
Description: 14 months old weighing 8.5 kg diagnosed to have PDA during vaccination. Examination revealed bounding pulses and continous murmur at left upper sternal border. Chest X ray revealed cardiomegaly and increased lung vascularity. After securing femoral venous access, PA and RA pressures were measured showing mean of 6 & 15 mm Hg respectively. After crossing PDA from venous side, 6 Fr delivery sheath was introduced but sheath was unable to negotiate suprarenal IVC due to severe venous spasm not relieved with saline push and Nitrate injections. While reattempting procedure contrast injection into femoral vein revealed duplication of infrarenal portion of IVC along with filling up of left paraspinal venous plexus causing decreased effective lumen of supra renal IVC in presence of normal RA pressure. Duplication was missed during the initial injection given high into one of the IVC lumen. After saline boluses and Nitrate injections 6/4 Duct Occluder was deployed successfully.
Conclusion: Venous spasm is more pronounced with low filling pressures. In presence of duplication of IVC with another venous egress effective lumen of IVC is reduced making it more prone for spasm even in presence of adequate filling pressure. Delineating IVC anatomy with contrast given into femoral vein becomes crucial to diagnose such spasm which was missed otherwise if injected



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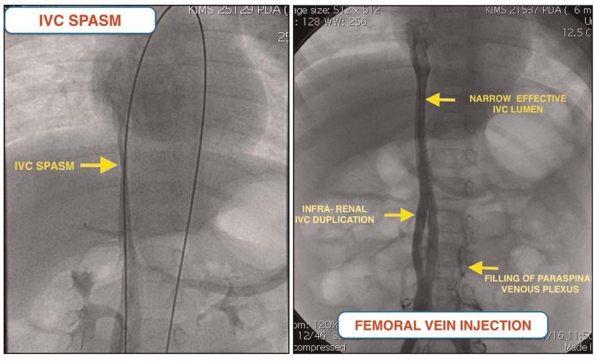

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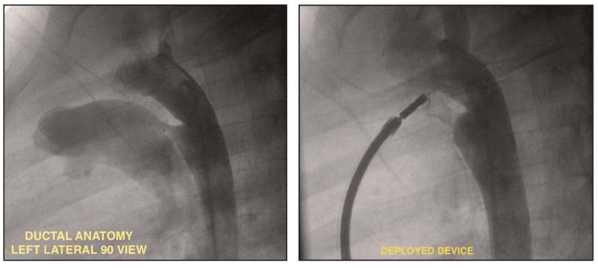

Figure 3.

P1928 - TANDEM PLACEMENT OF TWO DUCT OCCLUDERS GENERATION II DEVICES IN SUPEROINFERIOR VENTRICLES WITH CRISS CROSS ATRIOVENTRICULAR FLOWS

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Kims Hospital, Pediatric Cardiac Sciences, Hyderabad-India

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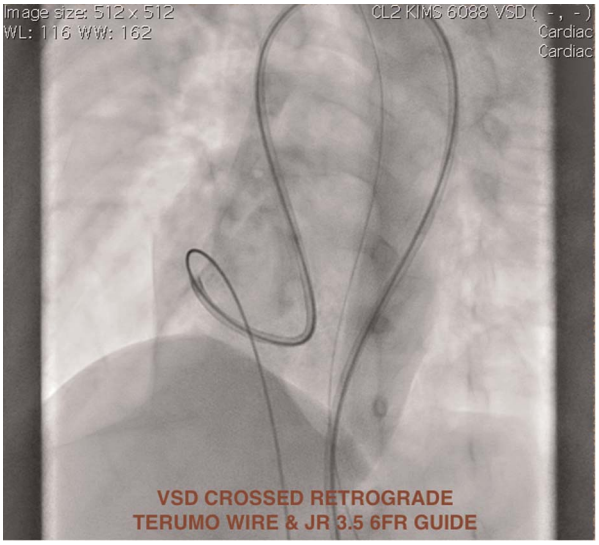
Introduction: Superior inferior ventricles with criss cross atrioventricular flows associated commonly with discordant Ventricular-arterial relationship. Here we are presenting a case of membranous septal aneurysm with multiple VSD in superoinferior ventricles and criss cross AV connection with AV - VA concordance, closed with tandem placement of two AMPLATZER DUCT OCCULDER II (ADO II) devices.

Case Details: 15 months old toddler weighing 7.2 kg presented with poor weight gain and recurrent respiratory tract infection. Referred with diagnosis of CCTGA and VSD. Examination revealed features of large left to right post tricupsid shunt with poor localisation of apex beat. Chest X-ray showed cardiomegaly, differential vascularity with right lung plethora. ECG revealed sinus rhythm, left superior QRS axis and biventricular forces. Echocardiogram revealed situs solitus, levocardia, juxtaposed right atrial appendage, superior inferior ventricles, AV-VA concordance, membranous septal aneurysm and multiple VSD's with moderate pulmonary artery hypertension. Oximetry run showed step up of 15% across right heart with QP/Qs of 2:1. PA pressure were 60% of the systemic pressure with mean of 35 mm Hg. Ventricular injections revealed superior position of RV and LV in inferior position with elongated LVOT. LPA was angulated with physiological oligemia due to streaming from MPA to RPA.Two ADO II devices (6/4 and 5/4 respectively) were deployed in tandem manner across septal aneurysm from retrograde technique with complete abolition of shunt and drop in PA pressure. In both cases VSD were crossed using 6Fr JR 3.5 guide catheter with 0.032 TERUMO ANGLED WIRE.



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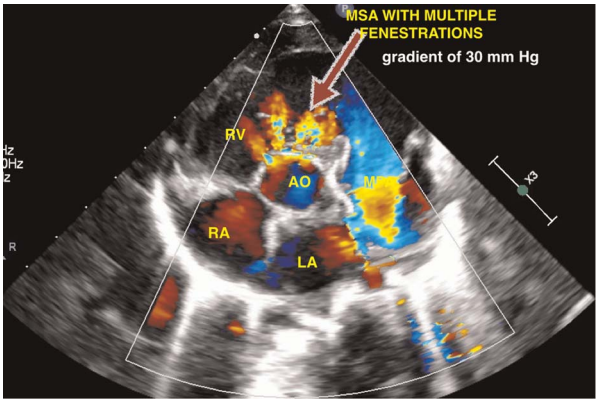

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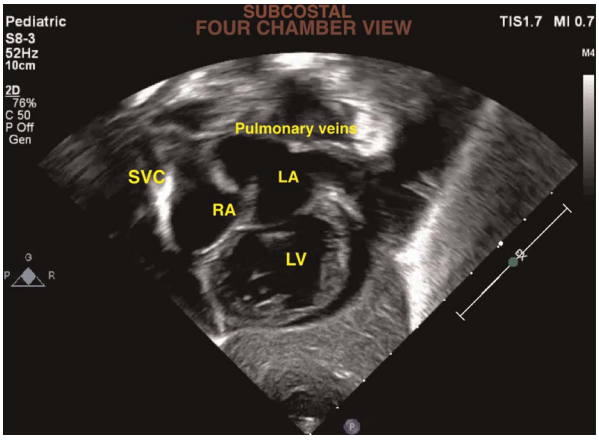

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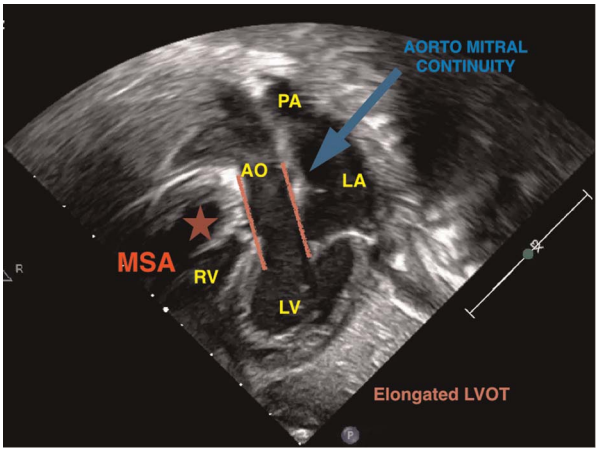

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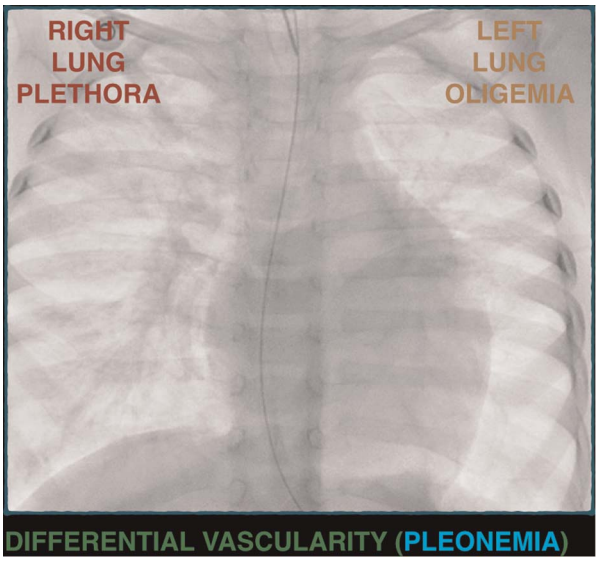

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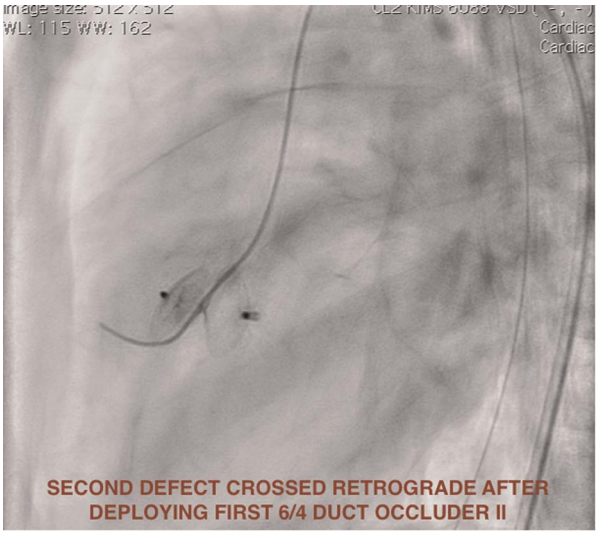

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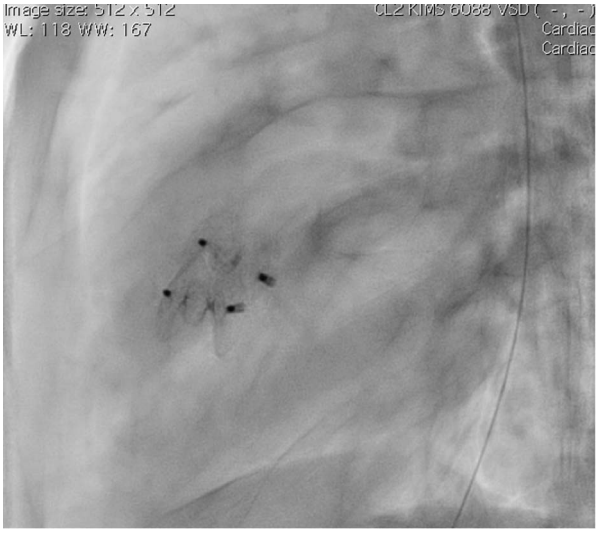

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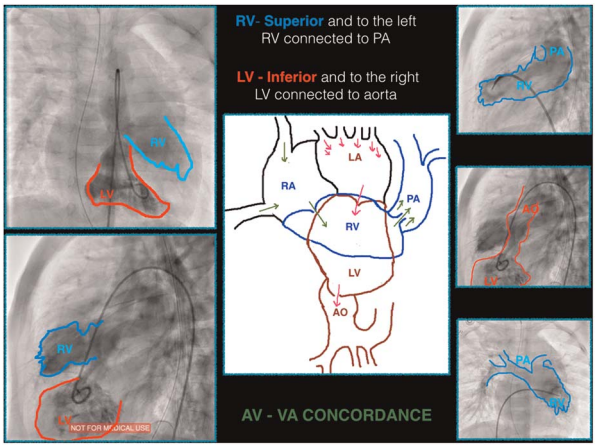

Figure 8.

Conclusion: Orientation of the IVS changes hence device closure is challenging in such cases. Delineation of the VSD by angiogram is difficult hence meticulous echocardiographic evaluation is needed. Tandem device placement with multiple ADO II is possible in such cases with membranous septal aneurysm and multiple defects.

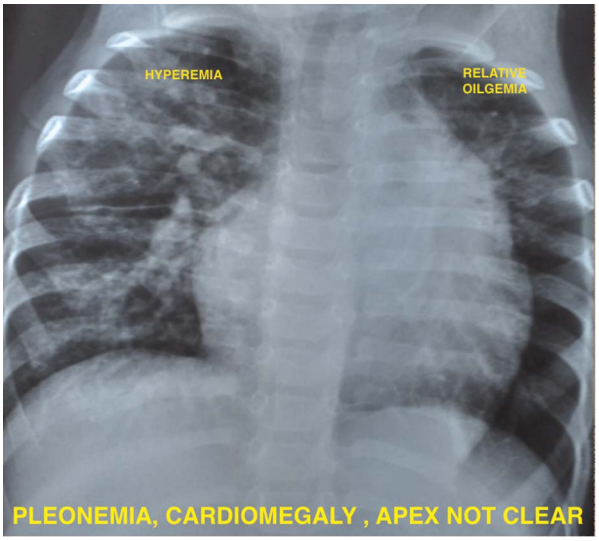

Figure 9.

P1958 - ASD WITH CYANOSIS EISENMENGER SYNDROME

Neeraj Awasthy[1], Dinesh Bhisht[2]
Max Hospital, Saket, Pediatric Cardiology, Delhi-India[1]; Max Hospital, Pediatric Cardiology, Delhi-India[2]

We Present A Series Of Cases Of Atrial Septal Defect Which Were Cynosed And Being Followed Up As Eisenmneger Syndrome. Case1: A 56 Year Old Female With Bronchiectasis Who Underwent Diagnostic Evlaution With Interesting Hemodynamics, Case2: 45 Year Case Of Connective Tissue Disorder, Case 3: With Lutembacher Syndrome, With Unroofed Coronary Sinus And Other Cases Have Been Enlisted. This Description Shows How A Case Of Asd With Cynosis Can Be Worked Up To Yield And Treat The Underlying Etiologies Of Pulmonary Artery Hypertension

P1960 - LV PSEUDOANEURYSM. PERCUTANEOUS CLOSURE

Neeraj Awasthy[1], Dinesh Bhisht[2]
Max Hospital, Pediatric Cardiology, Delhi-India[1]; Max Saket, Pediatric Cardiology, Delhi-India[2]

Percutaneous device closure of a left ventricular (LV) pseudoaneurysm have been rarely reported. We describe the case of a 60-year-old woman with a history closed mitral valvotomy at 18 years of age, Mitral Valve (MV) replacement at 53 year of age in Feb 2016 at 60 year of age. She presented with shortness of breath and chest pain since for 2 weeks. Echocardiography (Echo) revealed LV pseudoaneurysm with neck size of 12mm. Cardiac MRI showed a large LV pseudoaneurysm (36 × 32 mm) that was filling

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from a small leak in the anterolateral aspect of the ventricle. Considering high-risk candidate for surgical treatment in view of three previous sternotomies, the pseudoaneurysm was closed percutaneously with use of a 16-mm AMPLATZER muscular VSD occlude sucessfully. The patient was discharged from the hospital the next day and was asymptomatic on followup.

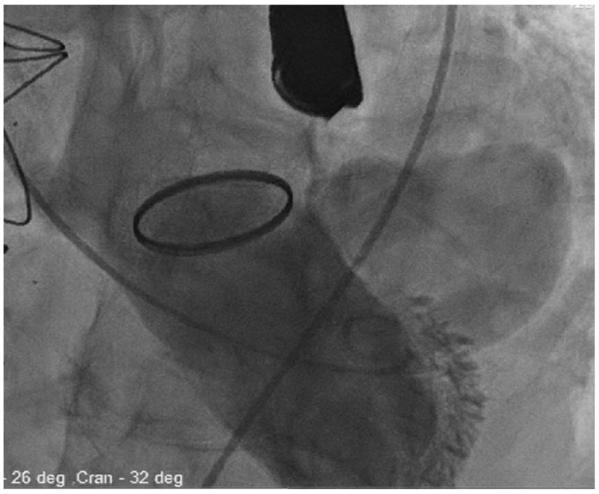

Figure 1.

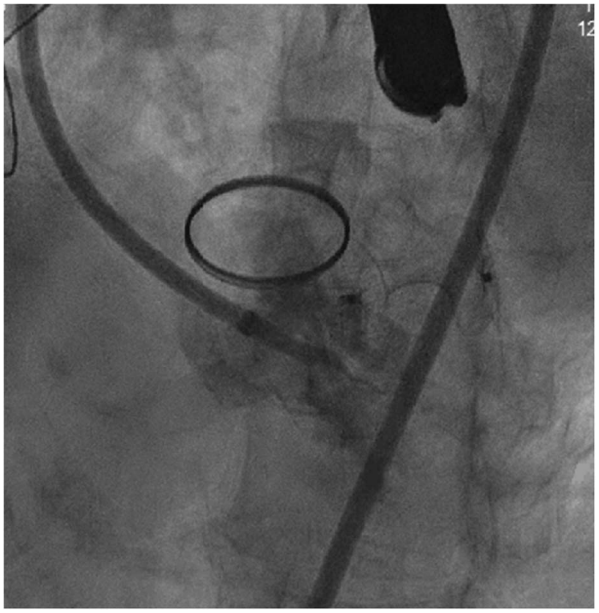

Figure 2.

P1968 - RETURNING BACK TO NATIV RIGHT VENTRICLE OUTFLOW

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Here in, we present a case with both native PA and non valved conduit that have been undergone closure of conduit due to RV enlargement after finding out the adequate native pulmonary outflow.

Case: A 24-years-old female who had been operated for double outlet right ventricule was admitted to our out-patient clinic for routine echocardiographic evaluation. She had undergone the Rastelli procedure at the age of four. Native PA reconstruction had not been performed due to the coronary anomaly. Transthoracic echocardiography showed enlarged RV, severe insufficiency in the nonvalved conduit. A detailed assessment revealed the presence of the native PA. At catheterization, RV injections showed two different outflows from the RV; one the native PA and the second the conduit (Fig. 1). By total occlusion of the conduit with sizing balloon, right ventricle systolic pressure rose from 40 to 90 mmHg. After establishing the adequate pulmonary annulus and valvular pulmonary stenosis, balloon pulmonary valvuloplasty was performed with a 24-mm balloon. RV pressure monitored again when the conduit was totally occluded with balloon. RV pressure remained unchanged about 40 mmHg. RV

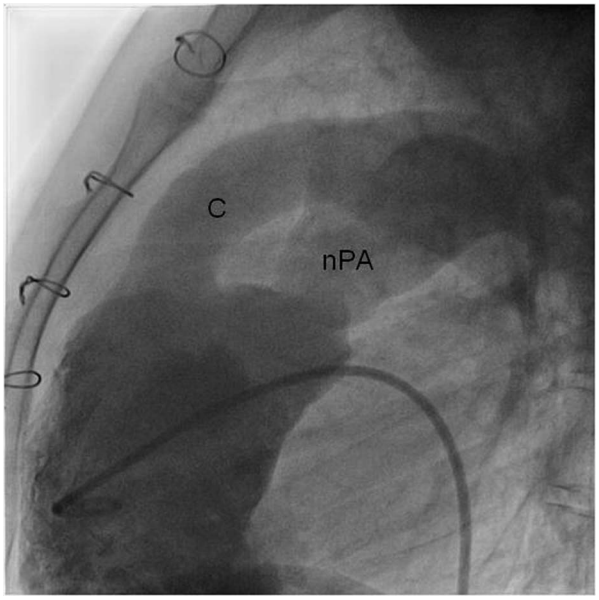

Figure 1.

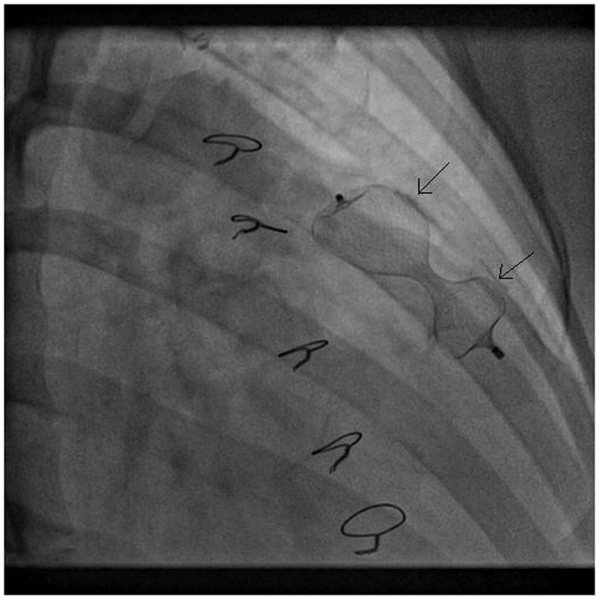

Figure 2.

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injection demonstrated the total occlusion of the conduit and RV decompression by native pulmonary artery. A 17 mm AmplatzerTM atrial septal defect occluder was deployed at the appropriate segment of the conduit (Fig 2). Post procedure echocardiography revealed mild degree of native PA stenosis and a residual forward flow in the conduit with mild degree of regurgitation. She did well without any complaints.
Conclusion: Finding out a native and adequate pulmonary outflow in a patient that undergone biventricular repair with a RV-PA conduit is an unexpected status. To our knowledge, this is the first case that right ventricle outflow redirected to native PA by transcatheter closure of the nonvalved RV-PA conduit.

P1998 - USEFULNESS AND SAFETY OF TRANSCATHETER CLOSURE OF ATRIAL SEPTAL DEFECT IN SMALL CHILDREN WEIGHTING 10KG OF LESS

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Background & Objectives: Transcatheter closure of atrial septal defect (ASD) has been accepted as a standard treatment. The risk-benefit relationship of transcatheter closure of ASD in the very young and small patients has not been clearly defined yet. Thus, to investigate the efficacy of safety of transcatheter closure of ASD in small children weighting less than 10 kg, we compared the data of patients with device closure and surgery.
Methods: From April 2004 to December 2015, 1149 ASD patients were treated with transcatheter closure. Among them, 124 patients weighting less than 10 kg were included as Device Group. During same periods, 350 patients were treated surgically, and among those 45 patients weighting less than 10 kg were included as Surgery group. Baseline characteristics, hemodynamic features, comorbidities, success rate and complication rate were compared between two groups.
Results: Although surgery was performed in younger, smaller children with relatively large defects, combined condition such as rim deficiency, multiple defects were not different between two groups. Except 4 cases (3.2%) of mitral encroachment in device group, all were successfully treated by either percutaneously or surgically. In both group, there was no major complication. Minor complication rates were significantly lower in device group than in surgical group (1.6% vs 8.8%, $p = 0.04$). Hospital stays were also significantly shorter in device group (4.0 ± 0.3 vs 11.1 ± 5.4, $p < 0.001$)
Conclusion: Transcatheter closure of secundum ASD with the ASO is technically feasible, safe and effective even in small children weighing less than 10 kg. We could suggest that even in very young children with ASD, no need to wait until they grow for transcatheter closure (>7.5 kg or age >1 year).

P2023 - TRANSCATHETER CLOSURE OF PATENT DUCTUS ARTERIOSUS IN VERY LOW BIRTH WEIGHT PRETERM INFANT FIRST REPORT IN THAILAND

Ratthapon Wongwandee, Boonchu Sirichongkolthong, Paweena Kaladee Thammasat, Pediatric (Cardiology), Bangkok-Thailand

Background: Transcatheter closure of Patent Ductus Arteriosus (PDA) under 2.5 kg preterm baby continues remain challenging due to difficult vascular access, variable duct diameter, temperature control and obstruction of adjacent vascular structures.
Method And Results: We report 2 preterm infants with procedural age 6, 12 days and procedural weight 2.4, 1.3 kg, respectively. Two babies had vascular access time 2, 4 minutes under ultrasound guided and procedure time 48, 58 minutes, respectively. Hemodynamic significant PDA(tubular type) were 3, 4 mm in mid diameter. Both of them used only venous access, 4 Fr JR catheter and Amplatzer Duct Occluder II additional size (ADO II AS, 04-04, 05-06) under echocardiogram and fluoroscopy. Successful device placement was achieved in 2 cases (100%). There were minor complications such as nonsignificant obstruction of left pulmonary artery and descending aorta, short period of platelet consumption in very low bithweight case (1.3 kg).
Conclusions: Transcatheter closure of PDA in very low bithweight infant remains challenging. We will develop technical skill to another choice for treatment of preterm PDA in our country.

P2056 - CARTO-ABLATION OF ATRIAL TACHYCARDIAS IN PRESCHOOL CHILDREN

Sergey Krivolapov[1], Roman Batalov[1], Liliya Svintsova[1], Olga Dzhaffarova[1], Irina Plotnikova[1], Sergey Popov[1], Igor Kovalev[2]
Cardiology Research Institute Pediatric cardiology department Tomsk-Russia[1]; Research and Clinical Institute for Pediatrics Pediatric cardiology department Moscow-Russia[2]

Importance: Prolonged X-ray radiation during RFA is a risk of remote complications especially in children of yearly age. Navigation mapping in such category of patients is perspective to reduce fluoroscopy time.
Aim: To assess the efficiency and safety of navigation mapping in RFA of atrial tachycardias in children.
Results: 30 RFA with CARTO system were performed to 23 children with atrial ectopic tachycardias during 2006 – 2016 years. The patients' age was 5,0 years (IQR: 3,58-6.0), ranging from 7 months to 7 years old. 7 patients were less than 15 kilos. The minimal age of the child who was performed CARTO was 7 months, the minimal weight – 7,4 kg.
As minimal size of the diagnostic catheter used in this system is 7 Fr (NaviStarTM), it is necessary to make a preliminary assessment of the diameter of femoral vessels by echocardiography.
Ectopic focus in most patients was in right atrium ($n = 20$). In two cases the approach to left atrium was performed by transseptal puncture, in one case – through patent foreman ovale.
The overall efficiency of CARTO ablation considering recurrences and retreatments was 91,3%. Vessel injuries and other complications, associated with a procedure, were absent. Due to CARTO ablation decrease of fluoroscopy time in 2,3 times in comparison with standard fluoroscopy mapping ($p < 0,001$) was marked.
Conclusion: RFA with navigation mapping is effective and safe in children weighing more than 7 kilos and allow reducing time of X-ray radiation to minimize complications associated with radiation exposure.

P2059 - RADIOFREQUENCY ABLATION OF TACHYARRHYTHMIAS IN CHILDREN TILL 5 YEARS OLD

Sergey Krivolapov[1], Roman Batalov[1], Sergey Popov[1], Liliya Svintsova[1], Igor Kovalev[2], Olga Dzhaffarova[1], Irina Plotnikova[1]
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88 patients under five years old underwent RFA. 22 of them were till one year old. Arrhythmias were WPW syndrome, atrial and ventricular tachycardias, AVNRT. Ablation was performed after failure of medication. Signs of arrhythmogenic cardiomyopathy had 68% of patients. One 5 Fr or 7Fr ablation catheter and one diagnostic catheter were for EP-study and RFA. Usually bifemoral access was used but in some cases we also used transesophageal reference catheter. In 20 patients with atrial ectopy we used non-fluoroscopic navigation system that allows minimizing radiation exposure. Access to the left side of the heart was performed through PFO, otherwise in 8 patients it was femoral artery puncture and in 3 patients it was atrial septal puncture. RFA was performed with 55 C with starting power 20 W, in case of conduction system damage risk a starting power was 5-10 W.
Results: First RFA procedure was effective in 78 patients (88,5%) but in 18 cases (23%) there was recurrence that was treated with 2nd procedure in all cases. There were still 3 cases of second recurrence in this group and third effective procedure was performed with good long term follow up. In 10 patients that was ineffective with first procedure in 6 cases there was a second procedure with good long term efficacy. The overall efficiency was 95%. Intraprocedural complications were transient AV-conduction disturbances in 4 patients. In one case with septal puncture there was asymptomatic hemopericardium. Reverse remodeling of arrhythmogenic cardiomyopathy in all patients.
Conclusions: RFA is safe and effective approach for difficult heart rhythm disorders management when medication fails. There is higher recurrence rate possibly due to minimally effective power applied. It is important to have echo and cardiac surgery back up for management of possible complications.

P2071 - A SYSTEMATIC REVIEW OF DEVICE CLOSURE OF PERIMEMBRANOUS VENTRICULAR SEPTAL DEFECT

Haripriya Santhanam[1], Linqi Yang[1], Zhaojin Chen[2], Swee Chye Quek[1]
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Background/Hypothesis: Device closure of ventricular septal defects (VSDs) is gaining popularity, although there remain concerns about adverse events; particularly heart block in perimembranous VSDs (pmVSDs). The aim of this study is to ascertain the outcomes (efficacy and complications) of device closure of pmVSDs, through a systematic review of currently available literature.
Materials and Methods: A PubMed and Scopus search for studies in English on device closure of pmVSDs published till mid-January 2017 was performed. Exclusion criteria included small series that had been included in multicenter studies, studies with sample size <5, and patients who had acquired VSD following myocardial infarction. The random effects model was used to obtain pooled estimates of success and complications.
Results: A total of 74 publications comprising 9165 patients with pmVSDs were included in this analysis. The age of patients ranged from 30 days to 73 years. The pooled estimate of successful device implantation was 97.3% (95% CI: 96.2%-98.2%). The most common complication was residual shunt (pooled estimated 10.9%; 95% CI: 7.2%-15.2%). Other complications included valvular defects (pooled estimate 3.8%; 95% CI: 2.4%-5.5%) and arrhythmias (pooled estimate 10.6%; 95% CI: 8.2%-13.2%). Of note, complete atrioventricular block (cAVB) occured in 1.5% (pooled estimated 0.8%; 95% CI: 0.4%-1.3%).
Conclusion: Our analysis suggests that device closure of pmVSDs is safe and yields good results. The complication of cAVB, while a concern especially with perimembranous type of VSD, is low (incidence = 1.5%). This is likely to be even more so with newer devices which are less stiff, and improved profiles. One limitation of the study is that different devices were used, and the results of each type could not be analyzed individually. Further studies validating this will be useful in formulating guidelines for the device closure of pmVSDs.

P2078 - CAN NUMERICAL SIMULATIONS SUPPORT PERCUTANEOUS PULMONARY VALVE IMPLANTATION FOR THE TREATMENT OF THE RIGHT VENTRICLE OUTFLOW DYSFUNCTION

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IRCCS Policlinico San Donato, Pediatric Cardiology and Adult Congenital Heart Disease, Milano-Italy[1]; Politecnico Di Milano, Bioengineering Department, Milano-Italy[2]; IRCCS Policlinico San Donato, Radiology, Milano-Italy[3]

Background: Percutaneous pulmonary valve implantation (PPVI) is a valid alternative to open-heart surgery for the treatment of right ventricle outflow tract (RVOT) dysfunction. However, an accurate pre-procedural evaluation of PPVI candidates is mandatory to minimize procedural complications (mostly arterial rupture, coronary compression, device embolization). For this purpose, we proposed a virtual simulation of PPVI implantation to assess the feasibility of the procedure and to elucidate potential PPVI-related risks before going in-vivo.
Methods: We enrolled five patients undergoing PPVI for right ventricle-to-pulmonary artery conduit failure. Each patient-specific anatomy, including the RVOT conduit, the calcifications, the aortic root and coronary arteries, was reconstructed from CT images using a commercial software (Materialise, Leuven-Belgium). Subsequently, the patient-specific computational model was complemented by the PPVI armamentarium, including the numerical model of both the stent and balloon as commercially available. Finally, a finite element analysis (FEA) was performed to reproduce the stent deployment in the RVOT after stent positioning and progressive balloon inflation, according to the real procedural PPVI standards.

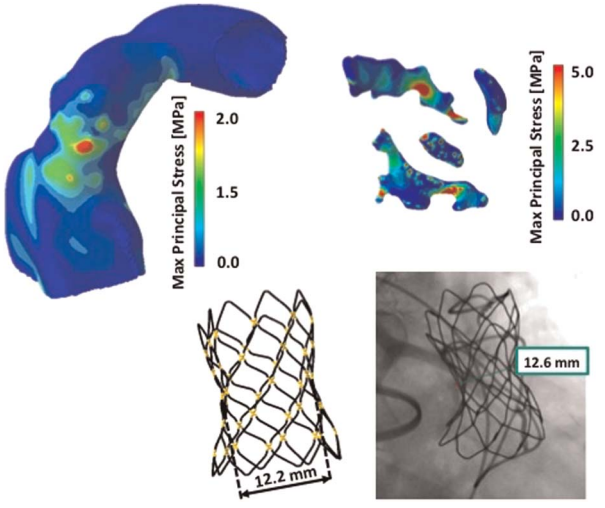

Figure 1.

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Results: The numerical approach proved able to virtually assess the anatomical RVOT changes after stent implantation in the single clinical scenario. Also, it quantified the interaction between the deployed stent and the anatomical RVOT structures pointing out anchoring forces, arterial wall stress, mechanical deformation of calcium, and risk of compression of the surrounding coronaries. Numerical results well compared to clinical evidences coming from the "in vivo" PPVI procedure performed in the Catheterization Suite: in-vivo diameters of the deployed stents were closed to what simulated, distances between the deployed stent and the left coronary were comparable, stent deformations occurred in the same locations. Figure 1.
Conclusions: Numerical methods can reliably reproduce the PPVI procedure and, in complex clinical-scenario, can effectively support the pre-procedural patient selection in order to minimize procedural complications.

P2099 - TRANSCATHETER CLOSURE OF AORTOPULMONARY WINDOW A CASE SERIES

Biswajit Bandyopadhyay[1], Amitabha Chattopadhyay[2], Mahua Roy[1], Saurabhi Das[1], Rhitajyoti Sengupta[3]
Rabindranath Tagore International Institute of Cardiac Sciences, Mukundapur, Kolkata, India., Pediatric Cardiology, Kolkata-India[1]; Narayana Superspeciality Hospital, Howrah, West Bengal,india, Pediatric Cardiology, Kolkata-India[2]; Rabindranath Tagore International Institute of Cardiac Sciences, Mukundapur, Kolkata, India., Pediatric Cardiac Intensive Care, Kolkata-India[3]

Background: Aortopulmonary window (AP window) is rare and early closure is recommended to prevent rapid development of pulmonary vascular obstructive disease. Transcatheter closure has low immediate and late complications, avoiding an extensive open heart surgery. We present our series of 9 children who underwent transcatheter closure.
Materials and Methods: 9 children (5 females, 4 males) between 1 to 6 years of age, underwent transcatheter closure of AP window over a period of 5 years (July 2011-August 2016) at our hospital. 8 had native lesions, one had a post operative residual lesion. The lesions were well defined echocardiographically from all views, contemplating the size and type of devices to be used along with estimation of the pulmonary arterial pressures. Oximetry run were done for all, before proceeding for the device closures. All had reversible PAH. Amplatzer Duct Occluder - I were used in 4, Atrial Septal Occluders in 2 and Amplatzer Duct Occluder II in 2. A muscular septal occluder was used for the post operative residual lesion.
Results: Per procedural transthoracic echocardiography were done for all to confirm the position of the devices. No residual leaks, aortic valve damage or coronary involvements were noted. One child with an ASD device had post intervention hematuria, which lasted for 3 days.7 had severe pulmonary arterial hypertension, which came down immediately after the procedure. One child had residual PAH post procedure, normalizing within 6 months. None of them had any complications on five years follow up.
Conclusion: Transcatheter closure of AP window in children is an effective and safe procedure with minimal complications. It should be considered in a favorable anatomy. Various devices may be used, the choice depending on the child's age and weight along with size of the defect, location and distance from the coronaries.

P2116 - MYOCARDIAL BRIDGING OF LEFT DESCENDING CORONARY ARTERY CAUSING MYOCARDIAL ISCHEMIA IN TWO BOYS

Sanja Dorner[1], Dragan Novosel[2]
University Hospital Osijek, Department of Pediatrics, Osijek-Croatia[1]; University Hospital Osijek, Department of Cardiology, Osijek-Croatia[2]

The authors present a two patients aged 12 and 14 who were admitted in the emergency room due to an anginal chest pain accompanied by nausea. The younger boy felt chest pain while he was running fast during football match in school. The older boy felt sharp chest pain after going out on cold winter day. Upon arrival, the both patients had normal clinical status, blood pressure and pulse rate, and no subjective problems. Recorded electrocardiograms were normal, but there were elevated levels of troponin I. Additional diagnostic procedures were done - in younger patient (12) echocardiography showed a structurally normal heart, no regional dyskinesia or valvular insufficiency. A MSCT coronarography showed myocardial bridging in the middle segment of the LAD. In the second patient (14) echocardiography revealed slight regional dyskinesia in apicoseptal region of the left ventricle. Coronary angiography performed at left anterior oblique projection showed narrowing in the middle LAD during systole, which returned to normal during diastole. Myocardial bridging (MB) is intramural course of the coronary arteries and a normal anatomic variant. The most common location is the middle part of the left descending coronary artery, but there was also described the involvement of the right coronary artery and left circumflex artery. The relevance of MB to clinical symptoms has been so far controversial. Though in the majority of cases isolated MB is a benign anomaly it may cause symptoms and in some cases may be a risk factor for sudden death. The challenges we are facing are to determine if the symptoms in symptomatic patients are caused by MB and to identify patients who are at increased risk for complication - especially a sudden death.

P2123 - STENT TREATMENT OF BRANCH PULMONARY ARTERY STENOSIS IN CHILDREN WITH CONGENITAL HEART DISEASE AFTER CARDIAC SURGICAL CORRECTION

Saule Kadirova[1], Frank Ing[2], Serik Alimbaev[1], Dmitriy Gorbunov[3], Almira Baygalkanova[4], Eric Otarbaev[1], Gulmura Momanova[5]
National Research Center For Cardiac Surgery, Interventional Cardiology, Astana-Kazakhstan[1]; Children's Hospital Los Angeles, Pediatric Cardiology, Los Angeles-United States[2]; National Research Center For Cardiac Surgery, Pediatric Cardiac Surgery, Astana-Kazakhstan[3]; National Research Center For Cardiac Surgery, Pediatric Cardiology, Astana-Kazakhstan[4]; National Research Center For Cardiac Surgery, Cardiac Catheterization Laboratory, Astana-Kazakhstan[5]

Background: Residual or recurrent branch pulmonary artery stenosis is a common finding after cardiac surgery for congenital heart disease. We report our experience using stents to treat branch pulmonary artery stenosis in children with congenital heart disease following surgical correction of stenotic or hypoplastic pulmonary arteries.
Materials and Methods: 22 children (15 males, 7 females) with post operative branch pulmonary artery stenosis were treated with stents between November 2014 to December 2016. Mean weight was $15{,}7 \pm 8{,}5$ kg (2,2 kg- 35 kg); mean age was $4{,}6 \pm 3{,}1$ years (1,5 months – 11 years). All patients had stenosis and/or hypoplasia of pulmonary artery branches associated with their congenital heart diseases prior to surgery. After surgical correction, stenosis developed at various areas along the pulmonary arterial tree including the ostium, and proximal branches. The stenoses varied from discrete or long segment and were either unilateral or bilateral.
Results: Stent implantation was successful for all 22 patients. 17 patients underwent transcatheter stent implantation of pulmonary branches and 5 patients received pulmonary artery stent implantation by a hybrid approach. Palmaz Genesis XD stents were used in 14 patients, Valeo Vascular stents in 7 and Genesis stent

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premounted on OPTA PRO in 1. After the procedure, the minimal diameter range of the pulmonary artery increased from 3,4 ± 1,6 mm to 8,8 ± 2,3 mm and the mean deviation of the right ventricular/pulmonary artery gradient decreased from 69 ± 10,4 mm Hg to 22 ± 4,3 mm.
Conclusion: Surgical correction of congenital heart disease and pulmonary artery stenosis can result in residual or recurrent stenosis requiring further treatment with stents. Our recent experience with this group of patients suggests that transcatheter and hybrid stenting of pulmonary artery branch stenosis is safe and effective.

P2135 - LEFT ATRIAL SIZE WILL INCREASE POST ASD DEVICE CLOSURE IN CHILDREN LESS THAN 3 YEARS OR LESS THAN 15 KG

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Magdi Yacoub Hospital, Pediatric Cardiology, Aswan-Egypt

Objectives: To evaluate how does the left atrial size change post atrial septal defect (ASD) occlusion in children ≤15 kg or less than 3 years old.
Background: small left atrial size is frequently encountered problem in patients with ASD specially in those who are ≤15 kg or less than 3 years old. We used the Amplatzer device in the ASD closure. The size of the left atrium in these patient had been followed up for 2 years post procedure.
Methods: Retrospective review of ASD procedures performed in children ≤15 kg or less than 3 years old.
Results: Between October 2009 and January 2017, 468 children underwent ASD Device closure. Amplatzer Device placement was successful in 440 of 468 patients. The number of patient who were less than 3 years old or less than 15 kg was 32. The weight of the patients ranged(11.6 ± 2.3 kg; P < 0.01) with Age of (2.5 ± 0.5 years P < 0.01). The left atrium transverse size was (1.9 ± 0.5 cm P < 0.01) which increased to (2.4 ± 0.3 cm P < 0.01) and longitudinal diameter of the left atrium was (2.5 ± 0.4 cm P < 0.001) which increased to (4.1 ± 0.6 cm P < 0.001). Major complications in all of the ASD device closure (n = 468) include 10 device embolization which retrieved percutenously apart from 2 retrieved by surgery, tamponad 1 patient required pericardiocentisis, complete heart block in 2 patients which improved after 2 weeks, supraventricular tachycardia in 3 patient one of them has CCTGA another 2 patients with AVNRT who required ablation post ASD closure.
Conclusions: The ASD device in children ≤15 kg or less than 3 years old is safe procedure. The LA size will increase with the follow-up although the left disc of the device seemed occupying most of it during the procedure.

P2146 - DEVICE INDUCED COARCTATION POST PDA CLOSURE

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Background: Patent ductus arteriosus (PDA) device transcatheter closure is a safe and effective procedure. Device induced coarctation is a well known complications which has been shown to be an avoidable complication.
Objectives: To evaluate that transcatheter closure with this device using different types of devices will show different incidence of device induced coarctation (DIC) .
Patients and Methods: Between January 2010 to January 2017, 489 patients (342 females and 147 males) with PDA underwent cardiac catheterization device closure using Amplatzer duct occluder (n = 293, 60%), PFM(PDA-R)occluder in (n = 161,33%) and coils (n = 35, 7%).
Results: The patient's age ranged from 3 months to 45 years (median 6 months). Successful PDA closure was achieved in 479 patients (97.9%) with 98% complete closure rate within 24 hours after the procedure. 10 patients (2.1%) had unsuccessful attempts, embolization of the device occurred in 5 of the patients (1.6%), encroachment of the aorta with angiography shown in 13% of the patient with no significant gradient (less than 30 mmHg) across the arch with invasive measurements before the release of the device. 10 patients out of the 489 had gradient of 30 mmHg at time of release of the device. Follow-up of the patients showed 5 patients developed significant obstruction of the arch post PDA device closure on which included 4 patients with Amplatzer device and 1 patient (all of which had been removed surgically)with one patient PDA-R, the obstruction was discovered on the follow-up between 6 month to 2 years. One of the patients who had surgical removal of the device developed recoarctation on the 6 month follow-up.
Conclusions: Duct device closure is safe and effective but carries mild risk of encroachment on the aortic arch which is minimally higher rate with Amplatzer Device compared to the PDA-R device.

P2174 - IMPLANTATION OF THE TRANSCATHETER HEART VALVE FOR PULMONARY POSITION IN NATIVE LARGE RIGHT VENTRICULAR OUTFLOW TRACT WITH SEVERE PULMONARY REGURGITATION

Alper Guzeltas, Ibrahim Cansaran Tanidir, Murat Sahin, Taner Kasar, Selman Gökalp, Hasan Candaş Kafali
Saglik Bilimleri University Istanbul Mehmet Akif Ersoy Thoracic and Cardiovasculer Surgery Training and Research Center Hospital, Pediatric Cardiology, Istanbul-Turkey

Background: We aimed to review the outcomes for the EDWARDS SAPIEN-XT® valve (Edwards Lifesciences, Irvine, California) for percutaneous pulmonary valve implantation (PPVI) in patients with native right ventricular outflow tract(RVOT).
Materials and Methods: Retrospective analysis was performed for all patients who have undergone PPVI using the EDWARDS SAPIEN-XT® valve with native RVOT.
Results: Five-teen patients (57% female, mean age 23 ± 10 years (range 12 to 46 years) were identified. Primary underlying diagnosis was tetralogy of Fallot (n = 13), Double outlet right ventricle (n = 1) and pulmonary stenosis (n = 1). Procedures were successful in all of the cases. In 3 cases (20%) pre-stenting was performed during the same procedure, and 6 or 10 weeks earlier in the rest of the cases. Among earlier stented patients, two cases had a second stent before PPVI because of stent fracture during the same procedure. Valve sizes were 26 mm (n = 2) and 29 mm (n = 13). Complications: one patient had moderate tricuspid regurgitation due to the procedure. There were no deaths during a median follow-up of 8.2 months (range 1 to 24 months). Valve function was preserved in all of the patients during follow-up.
Conclusions: The Edwards EDWARDS SAPIEN-XT® valve may be an alternative for patients with native large RVOT.

P2197 - CARDIAC CATHETERIZATION AND ANGIOGRAPHY IN THE NEWBORN PERIOD IN A SINGLE PEDIATRIC CARDIAC SURGERY CENTER

Alper Guzeltas[1], Gulhan Tunca Sahin[1], Ibrahim Cansaran Tanidir[1], Hasan Candas Kafali[1], Taner Kasar[1], Erkut Ozturk[1], Selman Gokalp[1], Akin Topkarci[2]

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Background: Nowadays, increasing number of diagnostic and interventional cardiac catheterization procedures can be performed during childhood even in the newborn period. Catheterization indications are usually different during newborn period because newborn patients are more prone to severe complications. We aimed to evaluate cardiac catheterization and angiography results and indications during newborn period.
Materials and Methods: Cardiac catheterization data were collected prospectively under Filemaker® program. Data of the newborn patients were retrospectively reviewed. Between January 2012 and November 2016, 2422 patients underwent cardiac catheterization, among them; 218 (9%) were newborn patients.
Results: One-hundred-twenty-five patients (57%) were male. The median age at cardiac catheterization was 7 days (1-30 days) and the median weight was 3 kg (1.8-4.2 kg). Diagnostic procedures were performed in 30 (14%) and interventional procedures were performed in 86% (n = 188) patients. Among interventional procedures stenting ductus arteriosus (n = 92), balloon pulmonary valvuloplasty (n = 35), balloon aortic valvuloplasty (n = 22), and balloon atrial septostomy/septoplasty were the most common procedures. Diagnostic cardiac catheterization revealed new or different anatomic information then echocardiography and cardiac CT in 6 patient. Median procedure time was 30 minutes (range 10-180 minutes), mean fluoroscopy time was 14.6 minutes (range 0-50 minutes). Mild to moderate complications (rhythm problems, femoral arterial spasm, respiratory/air way problems and bleeding) were observed in 40 patients (18%). Severe complications (cardiac tamponade in 3, and dead in 3) occurred in 6 patients (2.7%). Four patients underwent urgent surgical procedure.
Conclusions: Interventional procedures are more common during newborn period as an alternative for surgery or ameliorate the patient's condition before surgery. Surgery team should be aware of the patients undergoing interventional cardiac catheterization procedures in case of urgent surgical assistance requirement.

P2199 - THE HEMODYNAMIC EVALUATION OF SINGLE VENTRICLE PATIENTS WITH PULMONARY BANDING BEFORE BIDIRECTIONAL CAVOPULMONARY ANASTOMOSIS SINGLE CENTER EXPERIENCE

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Background: Our aim is to determine the risk factors for bidirectional cavopulmonary anastomosis (BCPA) in single ventricle patients with pulmonary banding.
Patients and Methods: 173 single ventricle patients who underwent diagnostic cardiac catheterization before BCPA were found at the hospital database search between January 2009 and December 2016. Out of 173 patients 34 patients who had pulmonary banding previously were found. The patient group was divided into 2 subgroups in accordance with their BCPA suitability at diagnostic cardiac catheterization. The data was analyzed to identify the risk factors of patients who were found unsuitable for BCPA.
Results: Twenty-five patients had banding of the main pulmonary artery (%74, MPAB), and 9 patients had bilateral pulmonary artery banding (%26, BPAB). All BPAB patients had concomittant aortic arch surgery, and 5 patients in total from both groups had atrial septectomy. There were no correlation between BCPA suitability and age at banding, age at diagnostic catheterization, ratio of pulmonary artery to aortic pressure (PAB/AOP) during banding, type of congenital heart defect, concomittant surgeries, type of banding (MPAB vs. BPAB). According to the method used to adjust band tightness (Trussler's formula vs. Adjustment just according to the pulmonary artery pressure and systemic O2 saturation), although not statistically significant the second method values was smaller in unsuitable patients. A positive correlation, between the age at catheterization and the PAP/AOP ratio for adjustment during banding was found.
Conclusion: Although older age at banding was reported as a risk factor in previous studies, no significant risk factors determined for BCPA suitability in the present study.

P2206 - TRANSCATHETER CLOSURE OF CORONARY FISTULAS EVENT FREE SURVIVAL AND SIZE OF THE FEEDING CORONARY ARTERY

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Background: During followup after transcatheter closure of coronary fistulas, ischemic events appear to occur more frequently after fistula closure at older age. It has been inferred that earlier closure may ameliorate the risk of such late complications. Supporting evidence for an underlying mechanism is lacking.
Methods: We performed a retrospective review of our experience with transcatheter closure of coronary fistulas during infancy and childhood with emphasis on event-free survival and size change of the feeding coronary artery. Fourteen patients underwent transcatheter closure of coronary fistulas at age 10 days to 18 years (weight 3.6-64.8 kg). The fistula originated from the right coronary in 5, from the left in 7, and from both in 2. There were no major procedural complications. Followup ranged from 1 month to 18 years.
Results: There were no ischemic events during followup. Recanalization of the fistula occurred in 1 patient and was successfully occluded in a second procedure. At the time of the procedure, the feeding coronary artery measured 3.2-9.0 mm in diameter by angiography and 3.6-8.3 mm by echocardiography. At latest followup, the echocardiographic diameter of the former feeding coronary measured 3.3-8.2 mm, suggesting only minimal regression in the size of the feeding vessel. The patient with recanalization had an increase in the size of the feeding coronary from 7.0 to 8.5 mm over 3 years. Our findings suggest that there is little regression in the size of the feeding coronary during followup but closure of the coronary fistula appears to prevent a size increase.
Conclusion: The benefit of coronary fistula closure at early age may be prevention of further coronary dilatation and allowing young patients to grow into the size of their feeding coronary vessel thus decreasing the risk of thrombotic events.

P2261 - AN AUDIT OF BALLOON AORTIC VALVOTOMY IN NEONATES WITH SEVERE AORTIC STENOSIS

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Background: There is no consensus among the international pediatric cardiac community about the optimal management of neonatal aortic stenosis. The choice of per-cutaneous versus surgical valvuloplasty remains one of an institutional preference. We reviewed the instituitional experience with neonatal balloon aortic valvuloplasty over a period of 6 years.
Methods: This was a retrospective case record analysis of all neonates who underwent a balloon aortic valvuloplasty at The Children's Hospital at Westmead from January 2011-November 2016.
Results: During the study period, 26 neonates underwent balloon aortic valvuloplasty. The median age at intervention was 4 days and the mean weight was 3.2 kg. 3 babies had moderate ventricular dysfunction and 6 had severe ventricular dysfunction. The valve morphology was bicuspid in 13 and unicuspid in 11. The median valve annulus was 6 mm (4.4-7.5 mm). The mean peak-to-peak gradient on cardiac catheterization was 52.5 mm Hg. Post procedure there was no or trivial aortic regurgitation in 10 patients, mild in 10 patients and moderate to severe in 6 patients. A higher balloon diameter to annulus ratio was associated with severe regurgitation ($P < 0.05$). There were 3 procedural complications. 1 neonate had an external iliac dissection and pulse loss that was managed conservatively and 2 suffered a peri-procedure cardiac arrest requiring resuscitation and extra-corporeal support. There were 16 re-interventions (11 in the same admission) with a median time to re-intervention of 21 days. There were 3 deaths in the group all of which were primarily attributed to the cardiac condition. There were no significant predictors for death and re-intervention in the group.
Conclusion: Balloon aortic valvotomy is a reasonable first line management for neonates with aortic stenosis. A high balloon diameter to annulus ratio is associated with an increased risk of significant aortic regurgitation.

P2262 - TRANSCATHETER CLOSURE IN PATIENTS WITH LARGE PATENT DUCTUS ARTERIOSUS AND MODERATE TO SEVERE PULMONARY HYPERTENSION A FOUR YEAR EXPERIENCE FROM NATIONAL REFERRAL CENTER IN INDONESIA

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Background: Trans-catheter closure has become a preferred procedure as compared to surgical ligation for the patent ductus arteriosus (PDA). However in large PDA with moderate to severe pulmonary hypertension, the studies are limited. This study aims to evaluate the efficacy and complications of this procedure.
Methods: Retrospective study has been conducted from January 2013 until December 2016 in National Cardiovascular Center Harapan Kita, Jakarta-Indonesia. Thirty three patients underwent trans-catheter closure. The inclusion criteria were PDA with diameter ≥8mm with recorded moderate to severe pulmonary hypertension mean pulmonary arterial pressure (mPAP) ≥ 40 mmHg by right heart catheterization). The results after procedure were observed clinically and by echocardiography.
Results: Median age was 19 years old (3-47 years old). Mean duct size was 9.8 (8.0-20)mm and with average mPAP was 58.7 ± 8.1 mmHg. There was no cardiac death. During the procedure, there was one case of—— device dislodge which underwent surgical ligation afterwards. Initial residual PDA before discharge was 60.6% which all comprises of minimum centrally residual. Upon follow up from one week to four months, no residual PDA was detected. A patient developed AV fistula after the procedure and underwent uneventful surgical ligation afterwards. The median length of stay in hospital was 3 (2-9) days.
Conclusions: Trans-catheter closure in large PDA with moderate to severe pulmonary hypertension was effective with minimum complication.

P2265 - SHORT TERM RESULTS OF PERCUTANEOUS BALLOON PULMONARY VALVULOPLASTY IN PULMONARY VALVE STENOSIS SINGLE CENTER EXPERIENCE

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Background: Percutaneous balloon pulmonary valvuloplasty (BPV) is the preferred interventional procedure for pulmonary valve (PS). The aim of this study was to evaluate the short-term efficacy of this procedure, from infant to adult, in our center.
Methods: This study included 62 patients with PS underwent BPV between January 2013 and December 2015 in National Cardiovascular Center Harapan Kita, Jakarta, Indonesia. Short-term efficacy such as residual trans-valvular gradient, pulmonary regurgitation, and complications were evaluated.
Results: Median age was 5 years old (range 1 month-40 years old). Median trans-pulmonary gradient before BPV, measured during the procedure, was 71 (range: 28-128) mmHg and 28.5 (range: 8-91) mmHg immediately after ($p < 0.001$). There was no cardiac death. Four patients developed acute limb ischemia, one patient developed total AV block and one patient developed acute pulmonary edema. Mild pulmonary regurgitation was observed in 33.9% and moderate pulmonary regurgitation occurred in 6.5% during one week follow after the procedure
Conclusions: Percutaneous balloon pulmonary valvuloplasty is an effective procedure in the treatment of pulmonary valve stenosis with good short-term results in our center.
Keywords: Pulmonary stenosis, balloon pulmonary valvuloplasty, short-term results

P2271 - BALLOON ATRIAL SEPTOSTOMY GUIDED BY ECHOCARDIOGRAPHY AT NATIONAL CARDIOVASCULAR CENTER HARAPAN KITA JAKARTA INDONESIA

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Background: Traditionally, balloon atrial septostomy (BAS) was carried out in catheterization laboratories assisted by fluoroscopy. During the 80 s, people found the advantages of echocardiography guiding during this procedure and performed at neonatal intensive care unit.
Methods: Subjects were all patients who underwent BAS at National Cardiovascular Center Harapan Kita Hospital during 2013 until 2015. The procedure was carried out either in the catheterization laboratory assited by fluoroscopy or in the intensive care unit guided by echocardiography.

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Results: There were 40 patients underwent BAS, but 7 patients were excluded due to lack of data. The median age during the procedure was 38 (range 1 to 433) days old. Twenty (60%) patients were diagnosed with transposition of the great arteries with intact ventricular septum. The remainder of patients was tricuspid atresia, pulmonary atresia with intact ventricular septum, and hypoplastic left heart syndrome. There were 8 patients (group A) who underwent BAS at catheterization laboratory and 25 patients (group B) at intensive care unit. No intra-procedural mortality was documented. There was no significant difference found between the 2 groups in age and hospital length of stay (90.7 + /- 159.9 vs. 19.8 + /- 27.3, p = 0.72, and 21.8 + /- 8.8 vs. 29.8 + /- 12.2, p = 0.13, respectively). No statistically difference was also found between the 2 groups in the difference of oxygen saturation level pre and post-procedure and the difference of inter-atrial septum defect diameter pre and post-procedure (30.5 + /- 23.3 vs. 29.7 + /- 14.7, p = 0.91, 6.0 + /- 1.5 vs. 5.8 + /- 1.7, p = 0.88, respectively).
Conclusion: Balloon atrial septostomy performed in the intensive care unit guided by echocardiography gives the same effectiveness and results compared to the one performed in the catheterization laboratory guided by fluoroscopy. This is very important to minimize the cost in developing country such as Indonesia with economic issues.

P2275 - NEW PERSPECTIVES IN CARDIAC SURGERY THE VIRTUAL VENTRICLE AND THE MOLECULAR CARDIAC SURGERY

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Purpose: Congenital Heart Surgery in complex malformations still carries high risk and poor outcome. Is there any new treatment on the light of the modern tools offered by the molecular biology?
Methods: We anatomically assessed the hypothesis that the cardiac ventriculo-arterial malformations in any morphological settings are sequential stages of the same embryogenetic and teratological process at ventricular level: the malrotation of the Trabecula Septomarginalis (TSM Leonardo's cord, Virtual Ventricle). We reviewed the current tools offered by the Molecular Biology with the enrichment strategies to amplify the targeted DNA and the potential of CRISPR/Cas9 technique in Congenital Heart malformations.
Results: 1- The TSM on the normal V shape or variants follows the development of the right ventricle what we refere as Virtual Ventricle. Te sequential TSM malrotation is an anatomical teratological continuum (Swalbe 1906) what we propose as Human Model for investigating the developing heart. 2- Each cardiac phenotype has a specific molecular expression and the CRISPR/Cas9 represent a very promising technique for interfering with the ongoing cardiogenesis.
Conclusions: The TSM malrotation model and the new techniques of molecular biology open to the possibility of not invasive early diagnosis in CHD and consequent molecular treatments during pregnancy what we refer as Molecular Cardiac Surgery.

P2277 - TRANSCATHETER CLOSURE OF VERY LARGE PATENT DUCTUS AARTERIOSUS (>1.0 CM) IN CHILDREN

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Background: Transcatheter treatment of very large PDA especially associated with pulmonary hypertension is challenging. Large PDA can associate with severe pulmonary hypertension and occlusion may result in hypertensive crisis, device embolization, aortic and left pulmonary artery obstruction and residual shunts.
Material and Methods: From August 2015 to July 2016, total 123 children with PDA were underwent transcatheter closure in Cardiac Medical Ward, Yankin Children Hospital, Yangon, Myanmar. Among these, seven patients, aged 7–13 years, with very large PDA and systemic or near systemic pulmonary arterial pressure were included.
Results: Successful device delivery and complete closure occurred in all patients (100% occlusion rate). Pulmonary arterial pressures were decreased significantly after closure. Follow-up evaluations

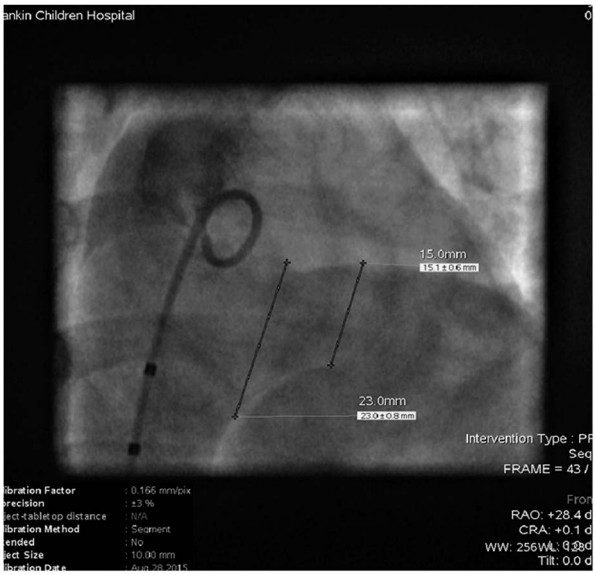

Figure 1.

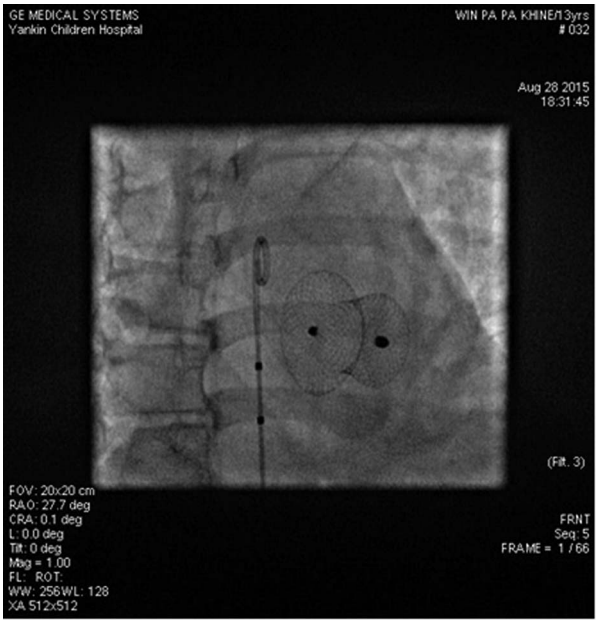

Figure 2.

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were made at 24 hours and 1, 6 months after closure. There were no complications.
Conclusions: Transcatheter Closure method is effective and successful in very large PDA associated with high pulmonary arterial pressure. Careful selection of device size and type is essential to avoid complications.
No 1 2 3 4 5 6 7
Age (yr) 13 7 10 12 10 12 8 Sex F F F F F F F
Body weight (Kg)30 18.0 19.0 35 25 19 15
Pul: end (mm) 15 11 12 14 17 14 12
PDA Type Conical Conical Conical Conical Conical Tubular Window
Used Device Lifetech Coccon Coccon Lifetech Lifetech Amplatzer Lifetech
PDA PDA PDA PDA PDA Muscular PDA
VSD
Device
Size(mm) 22 × 24 18 × 20 18 × 20 26 × 28 24 × 26 18 18 × 20

P2282 - Y STENTING IN COMPLEX PULMONARY ARTERY BIFURCATION STENOSIS

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Background: In complex pulmonary artery bifurcation stenosis (PBS) the distal pulmonary artery (PA) as well as the right and left PA ostia are involved. Literature on the effectiveness of different interventional strategies for PBS treatment is limited. We report on 11 children who were treated for complex PBS using the Y-stenting technique. Procedure data and long-term outcomes were analyzed.
Methods and Results: A stent is placed traversing the MPA to one PA branch. The telescope- and anchor technique are used to obtain access to the contralateral PA. By balloon dilatation the stent struts are opened after which a second stent is advanced through the stent struts into the contralateral PA. The second stent is placed with a 3-4 mm overlap with the MPA stent (clinched) or in the PA ostium (non-clinched). 11 Y-stenting procedures were performed, 9 in the main pulmonary bifurcation and 2 in the PA periphery. Control angiography showed unrestricted blood flow after all procedures. No major or minor adverse events were seen after stent implantation. 6 children needed re-intervention due to restenosis during median follow-up of 39 months. 4 of them received non-clinched bifurcation stents during the initial procedure.
Conclusion: Y-stenting was a safe and effective treatment in 11 children with complex PBS. It creates an artificial bifurcation close to the natural anatomy. Stenting sequence, the right materials and the telescope- and anchor technique are essential for a successful procedure. Due to high rates of restenosis in non-clinched stents, bifurcating stents should always be clinched.

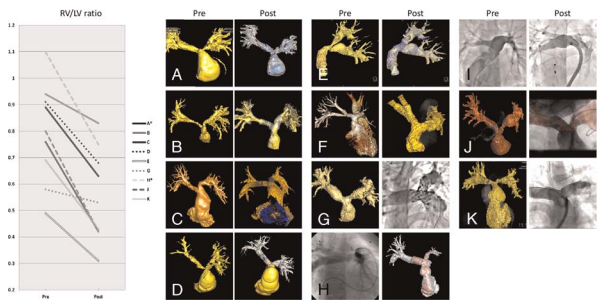

Figure.

P2303 - OUTCOME OF EMERGENCY INTERVENTIONS IN SICK NEONATES

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Introduction: The outcome of emergency interventions in sick neonates is not well established.
Methods: We retrospectively reviewed the records of sick neonates undergoing emergency cardiac intervention at our institute over a 7 year period. Neonates needing resuscitation, inotropes, prostaglandin or ventilatory support prior to the procedure were included. Neonates needing a procedure during non-working time of catheterization laboratory were also included.
Results: We identified 161 neonates who satisfied the inclusion and exclusion criteria for our study. Majority of neonates underwent an emergency balloon atrial septostomy (62.1%) with transposition of great arteries as the underlying lesion. Mean age of neonates in our study was around 10 days with mean change in saturation of 24% with BAS. 28 neonates needed an emergency aortic valve balloon dilatation, 11 needed a pulmonary valve balloon dilatation and 5 needed emergency coarctation of aorta dilatation. In all neonates AVBD or PVBD resulted in significant improvement in ventricular function. There were seven deaths out of 161 interventions done (3 in BAS, one each in AVBD, PV perforation, coarctation dilation, and PDA stenting). One of these neonates had B/L renal vein thrombosis with acute kidney injury and anuria. The local complication rate was also less than 5%.
Conclusion: Even emergency neonatal interventions are effective and rewarding in sicker neonates. However, they are associated with significant challenges. Hence, fetal diagnosis or early neonatal diagnosis and early referral should be emphasized.

P2308 - ATRIAL FLOW REGULATOR FOR LEFT HEART FAILURE A PILOT STUDY

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Background: Left ventricular diastolic dysfunction is commonly characterized by elevated left atrial pressure. Our limited study suggested that the reduction of LAP by creating a left-to-right interatrial shunt using a Atrial flow regulator resulted in significant reduction of LA pressure. This device helps in improving exercise tolerance and protecting from episodes of acute pulmonary edema.
Materials and Methods: our study includes 3 patients. One was a 67 yrs old male hypertensive post CABG with diastolic heart failure in class III NYHA,with episodes of Acute pulmonary edema. Second patient was a 13 yrs old girl with Restrictive cardiomyopathy in class III NYHA. Third patient was 35 yrs old DORV VSD, severe PAH, congenital MS with both precapillary and postcapillary hypertension in class III NYHA.
Results: First patient had a basal LA mean of 21, RA mean 8, he underwent interatrial device with AFR 8/5 mm device his post LA mean was 7. Second patient with restrictive cardiomyopathy had a LA pressure of 20 and reduced to 11 after 6/5 mm AFR device. Third patient had a RA mean of 13 mmhg and Basal LA mean was 17 mmhg, post AFR with a 10/5 mm device his RA and LA mean was 13mmhg Post procedure all the 3 patients had significant improvement of their symptoms. 2 patients are in class I NYHA, the 3 patient with severe PAH is in class II NYHA.
Conclusion: AFR regulates the rising LA pressures, thus improving exercise tolerance and protecting from episodes of acute

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pulmonary edema. This might lead to a reduction of hospitalizations, increase of quality of life and potentially diminish mortality in patients with diastolic heart failure. But further studies are needed to establish its effectiveness in long term follow up.

P2316 - COMBINED SEMILUNAR VALVE STENOSIS IN NEWBORN

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Background: Combined pulmonary and aortic valve stenosis is an extremely rare congenital heart anomaly. Neonates with this combination often present a life-threatening situation and require urgent treatment. The management approach for this cardiopathy is not well defined though percutaneous valvuloplasty seems to be the better option.
Case Report: We present the case of a newborn prenatally diagnosed with moderate pulmonary valve stenosis with intact ventricular septum. Postnatal echocardiographic assessment confirmed the presence of pulmonary valve stenosis and revealed the existence of associated aortic valve stenosis. Infusions of prostaglandin E1 and respiratory support with noninvasive mechanical ventilation were started immediately after birth. Patient remained stable and maintained a normal cardiac output. The intervention showed a severe combined semilunar valve stenosis and, accordingly, bilateral balloon valvuloplasty was performed. The procedure was successful. Pulmonary valvuloplasty was first performed, significantly reducing RV-PA gradient (from 77 to 25 mmHg) and slightly increasing LV-AO gradient (48 to 61 mmHg). Subsequent aortic valvuloplasty reduced LV-AO gradient to 27 mmHg. Patient was maintained in observation care status for 4 days and remained hemodynamically stable. Mild combined valve stenosis persisted as residual lesion.
Conclusion: Isolated pulmonary valve stenosis and isolated aortic valve stenosis are relatively common congenital heart diseases. However, combination of right and left ventricular outflow tracts obstruction is a very infrequent condition which warrants an early intervention. Very few cases are reported and there is not a universal agreement in the available literature on the approach to managing these patients. Nonetheless, the interventional transcutaneous approach is probably the best treatment option. Sequencing is controversial but it is probably better to start with the most affected valve (in our case the pulmonary one) considering that an increase of the transvalvular aortic gradient is very likely after the dilatation. It would be desirable to develop guidelines for sequential interventional therapy for this lesion.

P2319 - TRANSCATHETER CLOSURE OF MULTIPLE ATRIAL SEPTAL DEFECTS OUR EXPERIENCE

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Background: Transcatheter closure of multiple secundum ASD is technically challenging. We present our experience with double ASD device closures.
Materials and Methods: From May 2014- June 2016, 16 patients underwent transcatheter closure of multiple ASD defects in our Centre. Double device was considered only if the distance between two defects were more than 6 mm. Patients age ranged from 11 to 57 yrs with a mean of 37. The weight ranged between 30 kgs to 80 kgs with a mean of 58 kgs. The smallest ASD measured in TEE was 5mm and largest was 32 mm. Usually the two devices were positioned as a sandwich if both are larger devices but sometimes if both the devices are smaller the devices were interlaced .
Results: Totally 32 devices were deployed in 16 patients. The mean diameters of the larger and smaller defects were 30 and 9 mm. Balloon sizing was used in 10 patients. In 6 patients device was chosen only based on our TEE. The mean larger and smaller device diameters were 32 and 11 mm,.The largest device in a single patient was 32 mm and 20 mm, and smallest being 12 mm and 14mm.The mean Pre procedural PA pressure was 25mmhg and post mean PA pressure was 19 mmhg. Out of 14 patients 4 were snare assisted. Post procedure 12 patients had complete closure and 4 had residual shunts. At 1 month followup 2 patients had residual shunts, and at 6 months follow up none of them had residual shunts. All the patients were followed up on dual antiplatlets for 1 year.
Conclusion: Transcatheter closure of multiple ASD defects is a safe and reproducible technique. However appropriate patient selection and preprocedure TEE imaging play a vital role for better clinical outcome

P2329 - TRANSCATHETER VENTRICULAR SEPTAL DEFECT CLOSURE WITH PLATINUM COATED NITINOL DEVICE

Pornthep Lertsapcharoen[1], Suphot Srimahachota[2], Vidhavas La-Orkhun[1], Kanyalak Vithessonthi[1], Apichai Khongphatthanayothin[1]
Chulalongkorn University, Pediatrics, Bangkok-Thailand[1]; Chulalongkorn University, Medicine, Bangkok-Thailand[2]

Background: Transcatheter closure of perimembranous ventricular septal defect (pmVSD) remains a challenging issue due to the potential risk of heart block. The heart block mechanism is possible from too much stenting or radial force from the central waist of the device. A platinum-coated nitinol VSD device (Cocoon VSD device by Vascular Innovations Co Ltd, Thailand) was designed with the concept of soft central waist to minimize the risk of heart block.
Materials and Methods: During September 2011 to August 2016, 30 cases of pmVSD underwent transcatheter closure with Cocoon VSD device. The patients' ages ranged from 2 to 36 years, median of 7.5 years; and weights ranged from 10 to 86 kg, median of 30.2 kg. All patients had evidence of left ventricular volume overload and the Qp:Qs ranged from 1.3 to 2.8 with the mean of 1.8 + /-0.4.
Results: There were 28 successful device implantation cases with 2 failure cases. The 2 failure cases had surgical closure. Of the 28 successful cases, 4 cases developed hemolysis. Two hemolysis cases had device removal and VSD closure by surgery. Hemolysis resolved spontaneously in one case and after implantation of an additional ductal occluder in the other case. The complete closure rate was 57.7, 80.8 and 100% at 1 day, 3 month and 1 year, respectively. Within the mean follow-up period of 32.7 + /-11.8 months, there was no heart block in any cases. The hemolysis cases looked like to have too small size for the pmVSD diameter. The recommended size of the device should be at least 2 mm larger than that of left-ventricular-end pmVSD diameter.
Conclusions: Transcatheter pmVSD closure with platinum-coated nitinol device is safe and has a good closure result. With the property of soft central waist, it should minimize the risk of heart block.

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P2338 - HYBRID LARGE STENT IMPLANTATION FOR AORTIC RECOARCTATION IN AN INFANT

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Background/Hypotesis: Stent implantation is widely used to treat aortic coarctation. However, in small patients this procedure is limited by small size of peripheral arteries and growth concerns.
Materials and Methods: Report a novel hybrid procedure for implantation of large balloon expandable stent in an infant performed via right axillary thoracotomy
Results: A 9 kg 6-month-old infant with aortic coarctation underwent surgical repair at the age of 15-days and 45-days but remained with 60 mmHg transaortic gradient. He had had balloon aortoplasty but remained with transaortic pressure gradient (decresead from 60 to 35 mmHg). It was planned to use a stent that could be dilatable to adult size, by multiple dilatations during life. The novel hybrid procedure was performed in the cardiac catheterization laboratory under general anesthesia. The ascending aorta was achieved through a right axillary minithoracotomy, with a 3 cm horizontal skin incision between the anterior and posterior axillary lines, with a spare muscle technique, entering the chest in the third intercostal space. The patient received 100UI/Kg of heparin. A double suture purse string was done using a 5.0 polypropylene and a 7 F sheath was located into the ascending aorta under direct vision. Stent implantation was performed using a PALMAZ GENESIS 1910 stent mounted on 7 × 20mm POWERFLEX balloon. The stenosis waist disappeared at 6 atm and the pressure-gradient dropped from 60 to 0 mmHg. Good immediate angiographic, clinical, and hemodynamic results were obtained. There were no procedural complications. The patient was doing well at the 6-month follow-up. The transaortic pressure gradient remained low (15 mmHg by echo). Reintervention has not been required so far.
Conclusion: Large stent implantation through the ascending aorta via right axillary minithoracotomy, in a hybrid approach, to treat aortic recoarctation, was effective, enabling application of a large sheath and the implant of a re-expandable stent.

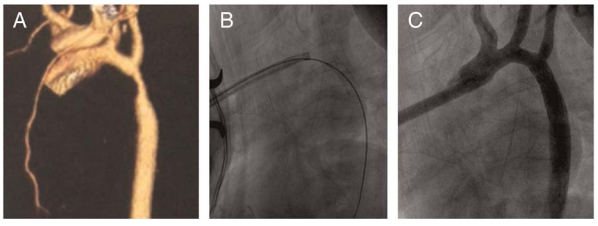

Figure.

P2345 - CATHCHAT AT RED CROSS CHILDREN'S HOSPITAL (RCWMCH) A NOVEL LIVE ONLINE TEACHING AND LEARNING PLATFORM FOR INTERVENTIONAL PAEDIATRIC CARDIAC CATHETERISATION

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Background: In Africa, paediatric cardiologists receive training in general cardiac catheterisation, but interventional skills can usually only be gained by undergoing fellowship training at overseas centres, or attending costly international interventional congresses. Consequently, there is a shortage of fully trained interventional paediatric cardiologists in Africa and only limited opportunities for such training exist. The incidence of paediatric heart disease is equal to, or may exceed that of developed countries and the majority of affected children will receive no treatment. There is a shortage of cardiac catheterisation laboratories in Africa, and an even greater, dire shortage of facilities for paediatric congenital heart surgery. Interventional cardiac catheterisation, however, holds promise to begin to address the epidemiological need, since surgical or intensive care facilities are usually not required for the interventional management of common, "simpler" congenital heart defects, e.g. ASDs, PDAs, and pulmonary stenosis.
Methods: At the RCWMCH, we have developed CATHCHAT, a system whereby interventional cardiac catheterisation procedures can be streamed live via the Internet. Local and international audiences log in to witness the procedures in real time and are able to interact directly with the operators in the cath lab. The online audience is able to follow the procedure in high-definition, step-by-step detail.
Results: CATHCHAT has been growing steadily over the past 3 years, and has broadcast more than 60 live cases to date. All paediatric cardiac centres in South Africa, as well as centres in Africa, Australia, India and Europe have viewed CATHCHAT, often simultaneously. The power of CATHCHAT is that renowned interventional experts can be invited to log in and advise the operators live while the procedure is performed. The logged-in audience may then witness the procedure, and learn from this interaction.
Conclusion: CATCHAT will stimulate the growth of paediatric interventional cardiac catheterisation capacity in Africa.

Figure.

P2346 - COMPLETE HEART BLOCK FOLLOWING TRANSCATHETER CLOSURE OF PERIMEMBRANOUS VSD USING AMPLATZER DUCT OCCLUDER II

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Trasnscatheter closure by device is now a well-established method of closure for perimembranous ventricular septal defect (PmVSD).

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One of the most serious complications of VSD device closure is complete atrioventricular block which has been reported in 3-18% is various studies following VSD device closure. Amplatzar Duct Occluder II (ADO-II) device is now commonly being used for closure of small to moderate sized PmVSD. Complete heart block has not been reported with ADO-II device. We are reporting 2 cases of complete heart block following transcatheter closure of PmVSD using ADO-II device where sinus rhythm was established following early surgical removal of the device. Case1: PmVSD device closure was done with 6/4 ADO-II device for a 8-year-old girl who developed CHB within 24 hours of procedure. She was started on intravenous steroid without any effect. Subsequently she was sent for surgical removal of VSD device and surgical closure of VSD after 48 hours of steroid therapy. She regined sinus rhythm after surgical removal of the device. Case 2: PmVSD device closure was done with a 6/4 ADO-II device for a 9-year-old boy who also developed CHB within 24 hours of VSD device closure. He was also started on intravenous steroid initially without any effect and subsequently sinus rhythm was regained after surgical removal of VSD device.
So CHB, though rare, can be seen after transcatheter closure of PmVSD using ADO-II device. This may be due to mechanical rubbing of the perinodal tissue by the discs of the device. It usually manifests early, within 24 hours of procedure. Late onset CAVB is very rare and has not been seen in our experience. Oral or intravenous steroids are having doubtful role in reversal of CAVB after VSD device closure. Early surgical removal of device prevents permanent damage of AV node and can restore sinus rhythm.

P2356 - PDA CLOSURE WITH CERAFLEX OCCLUDER IS THERE ANY ADDITIONAL BENEFIT

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Introduction: Although transcatheter closure of PDA is an established standard method, most frightening complication is protrusion of the aortic disc to the descending aorta (DAO) which may cause iatrogenic COA, especially in small children with small aorta. Ceraflex duct occluder (CDO) is a new device with similar properties with Amplatzer duct occluder (ADO). Device comes preassembled with the delivery cable by a loop connection through the holes and ready to load via the loader on the delivery cable. The loop made of surgical thread that provides the device to become flexible in 3600 directions.
Method: 21 patients underwent transcatheter closure with CDO.
Results: The median age was 1.2 years (6 months to 28 years) and weight was 9.6 kg (5.4-82). 11 patients were <1year and 11 had pulmonary hypertension. All had continuous murmur and all were type A. Narrowest PDA diameter was 4.1 ± 1.7 mm (2.2-8.2 mm). Intervention was successful in all. Final angiogram showed complete closure in 17/21 of them. Echocardiography achieved complete occlusion in all on the next day. There was no procedural complication.None of them showed evidence of stenosis at left pulmonary artery and DAO.
Discussion and Conclusion: Our results showed us CDO is a safe and efficacious in closure moderate to large PDA's whose duct morphology fit to the ADO. Its uniquely designed delivery system has an advantage in view of no applying tension to the device which provides the device in stable position and not changing the device position during and immediate after the releasing. It may give us an opportunity to be sure that the device not protrudes to the aorta after releasing, especially in infants those have small DAO.

P2361 - CORONARY BALLOON ASSISTED RETRIEVAL OF EMBOLISED VENTRICULAR SEPTAL OCCLUDER DEVICE

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History: An eight year old boy underwent cardiac surgery with intraventricular tunnel repair of the VSD and valve sparing Konno procedure for DORV with LVOT obstruction. Follow up echocardiogram showed multiple jets of residual VSD with largest measuring 5 mm with evidence of severe PAH. In view of sizeable VSD and significant PAH it was decided to close the largest of the residual VSD's. Device occlusion was preferred over surgical closure given the extensive patch and the PAH.
Imaging: After multiple attempts index residual VSD was crossed and 8 mm AGA muscular VSD device was deployed under fluoroscopic and TEE guidance. After confirming satisfactory position of the device on TEE, it was released. Unfortunately, approximately 15 minutes later the device embolized to RPA. Surgical retrieval was not immediately available and percutaneous retrieval was attempted. Despite multiple attempts with various catheters and usual hardware it was not possible to retrieve the device or even manipulate it into a favorable position. Thereafter, hard end of BHW coronary wire was introduced through the NITINOL mesh of the device and a 4 × 10 mm coronary balloon was placed over the wire. The coronary balloon was then inflated within the device and whole assembly was pulled across the pulmonary and the tricuspid valve up to the lower end of IVC. The device was then retrieved using bioptome through the 14 Fr sheath in right femoral vein.
Learning Point: Timely improvisation resulted in successful retrieval of embolized device in a child with difficult RVOT.

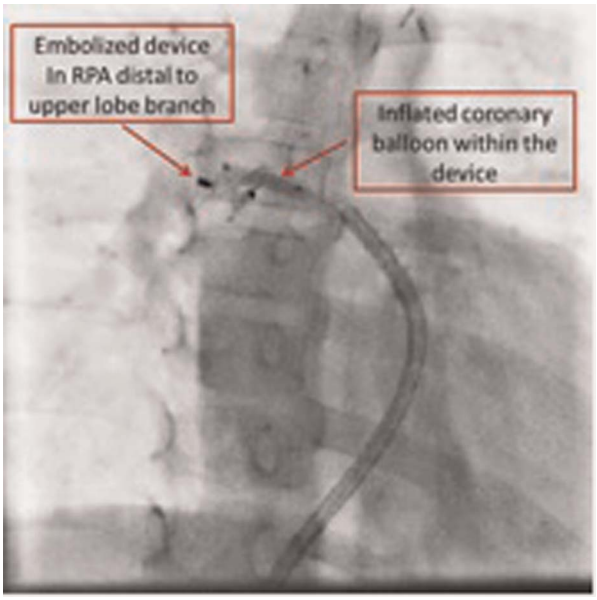

Figure.

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P2369 - TRANSCATHETER INTERVENTIONS AFTER GLENN ANASTOMOSIS AND FONTAN OPERATION IN PATIENTS WITH UNIVENTRICULAR HEART

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Introduction: We aimed to present transcatheter treatment of patients with a single ventricle physiology, experiencing low cardiac output (LCOS) or severe systemic desaturation (SSD).
Method: We retrospectively evaluated 33 patients between 2007 and 2016.
Results: The mean age was 7.6 years (6 months-21 years) and weight was 25.2 kg (6-54). The procedures were performed after a Kawashima, Glenn and Fontan surgery in 3, 12 and 18, respectively. SSD was encountered in 17. Closure of a Fontan fenestration was performed in 9. We occluded decompressing vein in 5 and pulmonary arteriovenous fistula in one. Closure of residual right SVC-atrium connection was performed in one and stent implantation to reroute the hepatic blood flow to the right lung in one, after Kawashima operation. The oxygen saturation of 79.3 8.1% increased to 92.25.6. LCOS and /or increased PA pressure was detected in the remaining 16. One was on an ECMO support. Amongst these 16, antegrade pulmonary flow (APF) was occluded using a number of devices in 7, APF was closed with the use of a covered stent, resolving associated left PA stenosis at the same time in two. Among 5 patients suffering from branch PA stenosis, 4 received stent implantation while the remaining was treated via cutting balloon angioplasty. Two separate stents were needed to treat branch PA and extracardiac conduit stenosis in one. In the patient on ECMO, fenestration was dilated with a balloon to ensure cardiac output. Fenestration was created in one. In patients with LCOS, the PA pressure decreased from mean 20.6 mmHg (15-27) to 14.9 mmHg (11-18). There was no procedural mortality. Circulatory failure regressed in all except one.
Conclusion: In the presence of LCOS or SSD, urgent catheterization should be considered. Significant PA stenosis should be treated even if there exists no pressure gradient throughout the circulation.

P2372 - ARTERIAL SEPTAL DEFECTS REPAIR IN CHILDREN BORN PREMTERM – LONG TIME FOLLOW UP

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Background and Hypothesis: One of the most common heart defects is atrial septal defect (ASD). The aim of this retrospective study was to asses long time follow up adverse events in preterm and term born children treated with ASD closure. We hypothesis increased adverse events among preterm children compared to term children due to the complex morbidity and cardiac remodelling in these children.
Material and Method: This is a retrospective cohort study including all children born in Sweden under the age of 18 years treated with percutaneous or surgical repair of ASD secundum between 2000 and 2014 at the Skåne University Hospital in Lund, Sweden. Demographic and adverse event data were retrieved from medical records and the Swedish National Birth Registry. Post-procedural complications occurring within short-term follow-up: ≤ 90 days and long-term follow-up: ≤365days was recorded and classified into minor and major events. Statistical analyses and Kaplan-Meier curve comparing differences and adverse events between preterm and term children were preformed.
Results: In total 344 children were included in the study; 269 term born children and 75 preterm born children. Preterm children were younger and lighter compared to term children at time of ASD closure. There was no difference according to method of ASD closure (device or surgical), the occurrence or the probability of occurrence of an adverse event between preterm and term children (Table 1). Further, there was no difference in

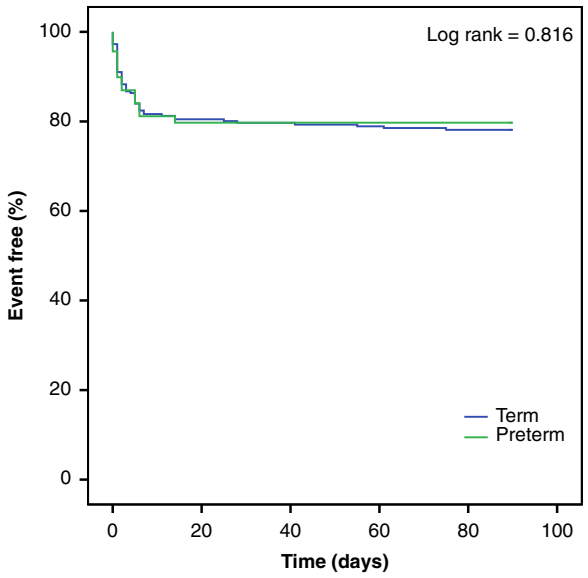

Figure 1.
Minor events within 90 days.

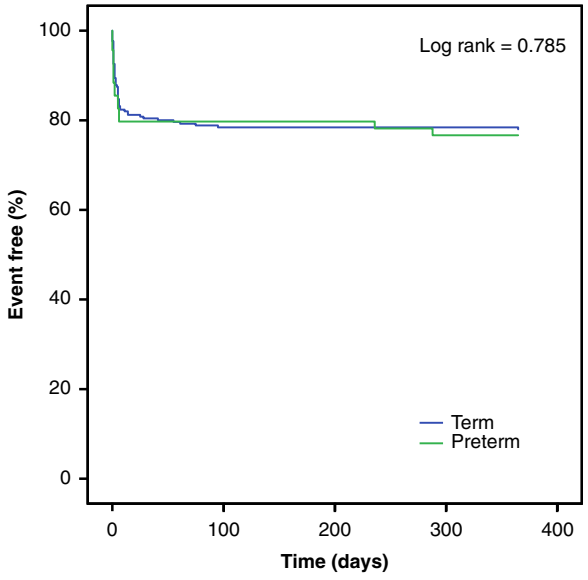

Figure 2.
Minor events within 365 days.

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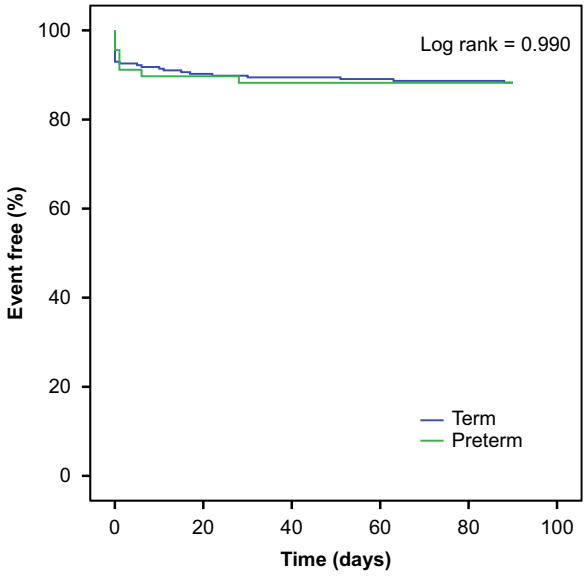

Figure 3.
Major events within 90 days.

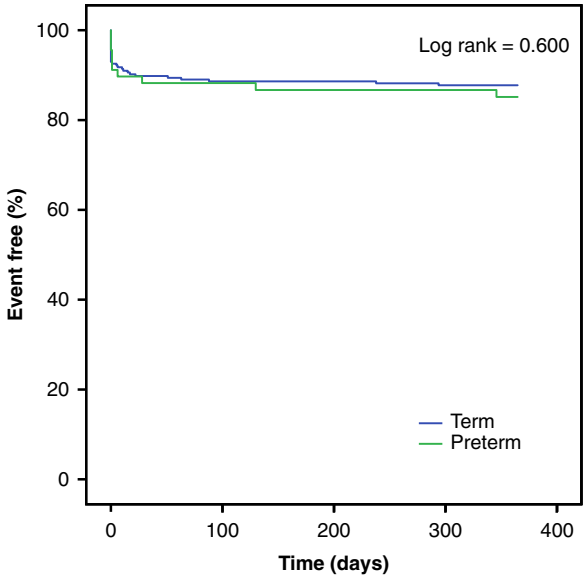

Figure 4.
Major events within 365 days.

Table 1. Information on study group and minor and major events.

	All (344)	Term (269)	Preterm(75)	*p*
Age (Years)	3.1(0.1-17.8)	3.4(0.1-17.8)	2.0(0.3-17.5)	<0.001°
Weight (kg)	14.5(3.5-110)	15.0(4.3-110)	11.5(3.5-65)	<0.001°
Device/Open	191 vs 153	150 vs 119	41 vs 34	0.886°°
Major events‡	38(12%)	30(12%)*	8(12%)**	0.922°°
Minor events‡	72(22%)	56(22%)*	14(20%)**	0,788°°
Major events‡‡	43(13%)	32(13%)*	11(16%)**	0.474°°
Minor events‡‡	73(23%)	57(22%)*	16(23%)**	0.9404°°

*Missing value 12 **Missing value 5 °Mann-Whitney U °°chi2 ‡Within 90 days
‡‡Within 365 days
Major: death, cardiac or respiratory arrest, stroke, device erosion, device embolization, permanent limb injury, emergent surgical, persitant arrhythmias, severe pericardial/pleural effusion, valvular insufficiency, vein obstruction, severe bleeding, pulmonary hypertension crisis, discharge >24 hours.
Minor: deployment malfunction, infection, minor bleeding, access site hematoma, prolonged limb paraesthesia, transient hypoxia during procedure, pericardial/pleural effusion, transient arrhythmia.

time to first event between preterm children and term children (Image1-4).
Conclusion: There is no difference in post-procedural adverse events between preterm and term born children, although the preterm born children were younger and less heavy at the time of ASD repair.

P2373 - TRANSCATHETER CLOSURE OF TUBULAR (TYPE C) PDA'S IN PATIENTS WITH PULMONARY HYPERTENSION

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Introduction: In this report, we will present our experience in transcatheter closure of Type C PDA's.
Method: Between 2005-2016, 898 patients underwent transcatheter PDA closure in our clinic. 41 of them had Type C PDA with pulmonary hypertension.
Results: The median age was 8 months (range 1 month to 6.5 years). The age groups were as follows: <1 year, 29/40 patients (73%); 1–2 years, 9/40 (22%) and >2 years 2/40 (5%). Median weight was 5.9 kg (range 3–15) with 53% of patients weighing less than 6 kg. The median measurements for minimal ductal diameter, length, and aortic ampulla were, 4.7 mm (3–11.7), 11 mm (4.3–24), and 7.7 mm (4–14), respectively. Pulmonary hypertension was found in all. PDA was closed with Duct occluder I (DOI) in 31, vascular plug (VP) 2 in five, VP I in two, VP4 in one, DO II in one and DO II AS in one. Procedure was successful in 37/40 (93%). Two patients sent to surgery after embolization of VP I and DOI. Significant aortic obstruction occurred before releasing the DOI device in another. Device displaced to descending aorta after 24 hours and repositioned with a bioptome in one. Occlusion rate was 100% after six month follow up. DOI slightly protrude to aortic lumen without significant obstruction in 13/31 infants. In these peak velocity at descending aorta decreased at follow up.
Conclusion: Although technically challenging, transcatheter closure of large Type C PDA especially in infants with various devices is possible. In a relatively small aorta, a larger aortic retention skirt diameter of DOI is one clinical concern; however, the somatic growth of aorta allows unimpeded aortic flows on late follow-up. In order to prevent aortic obstruction, plugs can be used to occlude the long Type C PDA especially in infants.

P2374 - TETRASOMY 5P MOSAICISM AND MULTIPLE CONGENITAL HEART DEFECTS

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Background: Tetrasomy 5p mosaicism is a rare genetic anomaly. There is a paucity of information on the relationship of Tetrasomy 5p mosaicism and associated congenital heart defects. We report the first case of a large atrial septal defect and other associated congenital cardiac anomalies in a patient affected with Tetrasomy 5p mosaicism.
Materials and Methods: A 2 year, 9 month old female was referred by pediatric genetics for evaluation. Her mother reported that the pregnancy was complicated by polyhydramnios and the patients past medical history was complicated by failure to thrive, hypotonia and severe scoliosis. She had not previously been referred or evaluated by a pediatric cardiologist despite an extensive medical evaluation at multiple outside institutions.

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Results: Her initial echocardiogram was significant for a large secundum atrial septal defect (ASD), moderately enlarged right atrium, moderately dilated right ventricle, tricuspid valve dysplasia, mild-moderate tricuspid valve regurgitation, estimated right ventricular systolic pressure of 50 mmHg, mitral valve dysplasia, mild mitral valve regurgitation and a small patent ductus arteriosus (PDA). A cardiac catheterization was performed and hemodynamics showed Qp/Qs = 4:1, indicating significant left to right shunting. Device closure of the ASD and PDA were not attempted because of concern for increased mitral valve regurgitation noted on a simultaneous transesophageal echocardiogram. Therefore she underwent subsequent surgical ASD patch closure and PDA ligation. Skin biopsy performed at the time of cardiac surgery was positive for Tetrasomy 5p mosaicism. The patient was discharged without any reportable concerns and continues to be followed for her tricuspid and mitral valve disease.
Conclusion: Our patient demonstrates previously unreported presence of multiple congenital heart defects in a child with Tetrasomy 5p mosaicism. If Tetrasomy 5p mosaicism is suspected the patient should be referred to a pediatric cardiologist and an echocardiogram performed.

P2376 - DIAGNOSTIC AND INTERVENTIONAL CARDIAC CATHETERIZATION IN CHILDREN SUPPORTED BY EXTRACORPOREAL MEMBRANE OXYGENATION A TEN YEAR SINGLE INSTITUTION REVIEW

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Background: Cardiac catheterization plays an important role in the management of children on extracorporeal membrane oxygenation (ECMO). We report our experience with diagnostic and interventional cardiac catheterization of children on ECMO.
Methods: A retrospective review of all children who underwent cardiac catheterization on ECMO at a single institution from January 1, 2007 through December 31, 2016.
Results: 41 catheterizations were performed on 35 patients (median weight 3.45 kg; range 2.0-63) within 3.1 ± 3.2 days of ECMO cannulation. 35/41 (85.3%) were performed in patients <1 year of age (21 < 1 month). 27 were performed in patients with congenital heart disease (24 within 30 days of cardiac surgery), 6 with cardiomyopathy/myocarditis, 6 post heart transplant, and 2 with non-cardiac diagnosis. 7 catheterizations revealed data that prompted surgical intervention. 28 cases had 31 interventions performed including: Transeptal puncture(3), Atrial septostomy/septoplasty (11), ASD closure(1), endomyocardial biopsy(1), Balloon angioplasty of aortic arch(1), coronary artery(1), or branch pulmonary artery(1), and stent placement in BT shunt(7), branch pulmonary arteries(4), right ventricle to pulmonary artery conduit(1), and Fontan fenestration(1). 17/35 patients survived to hospital discharge, including 3 who had heart transplant after ECMO. One patient required mediastinal exploration for bleeding. 2 patients had new onset seizures within 24 hours of catheterization, both of whom had their catheterization within 24 hours of going on ECMO. There were no other major adverse events, no vascular access complications, and no complications related to patient transport.
Conclusion: Cardiac catheterization with complex interventions can be performed safely in children supported by ECMO including smaller and post operative patients. This approach plays an important role in the diagnosis and treatment of hemodynamic and structural abnormalities in these patients.

P2381 - SAFETY AND EFFICACY OF PERCUTANEOUS CLOSURE OF PERIMEMBRANOUS VENTRICULAR SEPTAL DEFECTS IN CHILDREN IN A LIMITED RESOURCE SETTING IN SOUTH AFRICA

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Background/Hypothesis: Ventricular septal defect (VSD) is the most common congenital cardiac lesion. Surgical closure is the gold standard but in an isolated perimembranous ventricular septal defect (PMVSD) percutaneous closure is an attractive alternative, particularly in a limited resource setting. Our experience suggests that percutaneous closure of a perimembranous VSD, in the appropriately selected patient, is safe and effective.
Materials and methods: We performed a retrospective chart review of all children that underwent percutaneous closure of a PMVSD at Inkosi Albert Luthuli Central Hospital from October 2010 until December 2016. Patients that had percutaneous closure of any VSD other than PMVSD, including residual VSD post-surgical closure were excluded.
Results: Fourty-two patients were included in our retrospective analysis, 27 females and 15 males with a mean age of 6 years 6 months (Range: 2 years 9 months to 15 years 9 months). Mean follow up was 2 years 3 months. A total of 30 patients (71.4%) had complete closure of their defect. Eleven (26.2%) patients had a residual but haemodynamically insignificant defect and only 1 patient (2.4%) required surgical intervention due to failed percutaneous closure. Two patients had mild aortic regurgitation post procedure. Significant complications included 1 patient with moderate tricuspid regurgitation and 2 patients with device embolisation. In one of these patients, the embolised device was retrieved and replaced with a larger device. In the second patient, surgical retrieval and closure was required. No cases of heart block were recorded.
Conclusions: In our experience percutaneous closure of a perimembranous ventricular septal defect in a child appears to be safe and effective.

P2397 - HEALING OLD BROKEN HEARTS – THE EFFECT OF AGE ON THE RECOVERY FROM PERCUTANEOUS ATRIAL SEPTAL DEFECT (ASD) CLOSURE

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Hospital Sta Cruz, Pediatric Cardiology, Lisbon-Portugal[1]; Nagano Childrens Hospital, Pediatric Cardiology, Matsumoto-Japan[2]

Background: Percutaneous ASD closure is performed to both adults and children with hemodynamically significant ASDs. Right heart reverse remodeling has been demonstrated, though no study to our knowledge has looked at the differences according to age.
Materials and Methods: Retrospective study of 12 patients submitted to percutaneous ASD closure at a single center (6 <7 years and 6 >39 years). Echocardiographic 3D and strain data was acquired at fixed time points before and up to 2years after ASD closure.

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Longitudinal data analysis was performed with a linear mixed effects model.
Results: At baseline there was no statistically significant difference between age groups in terms of Qp/Qs (mean 1,92) and right ventricle end-diastolic volume (RVEDV) z score (mean +3,05). A tendency towards bigger right atria (RA, mean volume z score of 1,13 vs -0,34 p 0,04) and smaller left atria (LA, mean volume z score of -2,33 vs -1,42 p 0,04) was found in the adult population, while distensibility was not different at baseline. After repair, RV reverse remodeling was observed over time in both groups (mean RVEDV at 1 year -0,96, time term p value 0,001) without significant difference, with a tendency for steady lower RV ejection fraction in the adults (p 0,06). LA distensibility was lower through time in the adult population in both volumetric (p <0,001) and speckle tracking (p < 0,001) techniques, displaying a tendency to further diminish over time (group.time interaction p value: volumetric method 0,013; speckle tracking 0,098).
Conclusion: In our dataset both children and adult patients benefited equally from ASD closure concerning RV remodeling. However, LA distensibility showed a tendency to progressive impairment in the adult population. While further studies are needed to assert the clinical implications of these findings, this raises concerns about triggering of atrial arrhythmias.

P2402 - IMPLANTATION OF STENTS FOR POSTSURGICAL RECOARCTATION OF THE AORTA IN ADOLESCENTS AND ADULTS

Luiza Zalewska[1], Jacek Bialkowski[1], Malgorzata Szkutnik[1], Roland Fiszer[1], Szymon Pawlak[2], Beata Chodor[1]
Medical University of Silesia, Silesian Center For Heart Diseases, Congenital Heart Diseases and Pediatric Cardiology Department, Zabrze-Poland[1]; Medical University of Silesia, Silesian Center For Heart Diseases, Pediatric Cardiac Surgery Department, Zabrze-Poland[2]

Background: Results of stent implantation (SI) of postsurgical recoarctation of the aorta (ReCoA) are not frequently published. This study sought to retrospectively evaluate results of SI in ReCoA in older children and adults.
Material and Methods: Twenty-eight SI were performed on 26 ReCoA patients with a median age of 23 (10-65) years. Dependent upon availability, the following stents were applied - Palmaz, Cheatham Platinum (CP), Andrastents XL/XXL(AS), Covered CP (CVCP) stents and selfexpanding stents (Smart). Generally high pressure balloons were applied to dilate stents.
Results: The procedure was effective in 20/26 patients (77,7%). The mean gradient reduced from 40,5 + /- 18,7 mmHg to 13,1 + /- 12,1 mm Hg (p < 0,05) and the stenosed segment increased from 7,5 + /- 3.02 mm to 13,1 + /- 3,32 mm (p < 0,05). In 6 cases (including a patient treated with a Smart stent) transaortic gradient after SI remained >20 mm Hg (stiff postsurgical lesion). For one patient (40 year old male), an acute dissection of the aorta occurred during balloon predilatation. Immediate CVCP implantation resolved this problem. Two more CVCP were used – one to close a small aortic aneurysm which appeared 5 years after a Palmaz SI and one other to stabilize a broken CP bare metal stent. There were no deaths nor the aortic dissection during follow-up and most patients were able to reduce or suspend their medication for systemic hypertension.
Conclusions: Endovascular stenting of recoarctation of the aorta in adults and adolescents appears to be good a method of treatment in experienced hands. However, for some patients the presence of a stiff lesion can provoke suboptimal results. Considering the serious complications which may occur after SI, following surgery all patients should have regular follow up (including an imaging study). Covered stents should always be available in the cathlab as rescue device when implanting stents in CoA patients.

P2423 - PLANNING OF INTERVENTIONAL CORRECTION OF SINUS VENOSUS DEFECT WITH PARTIAL ANOMALOUS PULMONARY VENOUS DRAINAGE USING 3D PRINTING TECHNIQUES

Mari Nieves Velasco Forte[1], Nick Byrne[2], Antony Hermuzi[3], Gorka Gomez Ciriza[4], Pimpak Prachasilchai[3], Gur Mainzer[3], Kuberan Pushparajah[3], Tom Zellers[5], Michael Luna[5], Tarique Hussain[5], Israel Valverde[6], Shakeel Qureshi[3], Eric Rosenthan[3]
King, Division of Imaging Sciences And Cardiovascular Engineering, London-United Kingdom[1]; Guy, Medical Physics, London-United Kingdom[2]; Evelina Children, Paediatric Cardiology, London-United Kingdom[3]; Institute of Biomedicine, Cardiovascular Physiopathology, Seville-Spain[4]; UT Southwestern Medical Center, Paediatric Cardiology, Dallas-United States[5]; Institute of Biomedicine, Paediatric Cardiology, Seville-Spain[6]

Background: Sinus Venosus Defects (SVD) with anomalous pulmonary venous drainage (PAPVD) have traditionally been treated surgically. Cardiac magnetic resonance (CMR) is often used for confirmation of diagnosis and procedure planning. The introduction of a novel interventional approach was supported by 3D-printing techniques.
Methods: 3 patients (males, 74-128 kg, 30-66 years) diagnosed with SVD and PAPVD (right upper pulmonary vein to superior vena cava (SVC)) underwent CMR as part of our institutional protocol. A patient-specific 3D model was printed for each patient (figure1).

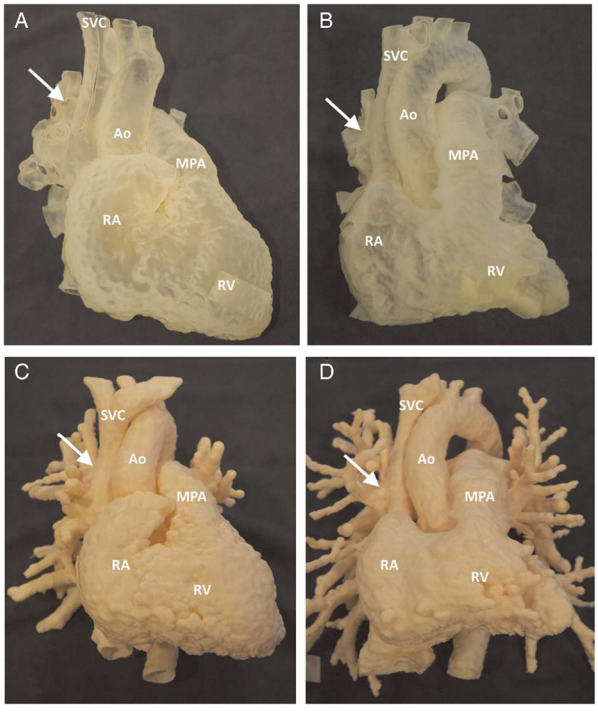

Figure 1.

https://doi.org/10.1017/S104795111700110X Published online by Cambridge University Press

Whole heart anatomy was segmented from 3D balance steady-state free precession images in two patients and time-resolved angiography in one, using Mimics Medical software (version 18.0). All models were printed in TangoPlus. A second polyurethane replica was printed for two patients. A stent was inserted in the SVC of the model to assess whether SVD closure and PAPVD flow redirection to left atrium (LA) could be achieved with a catheter based approach.
Results: CMR verified the diagnosis for all patients. Model based phantom stent insertion confirmed SVD closure and patency of the RUPV to the LA(figure 2). Measurements from the model and CMR were obtained for stent size. The technique was subsequently applied in the patients. A sizing balloon was used to size the SVC and occlude the defect. Simultaneously, a catheter was placed in the RUPV demonstrating patency of PAPVD, as previously shown in the model. This was also confirmed with transoesophageal echocardiography (TOE) and rotational angiography. Custom made covered Cheatham-Platinum stents were then mounted on a 28mm (2patients) and 26 × 60mm balloon. Once implanted, TOE and angiography confirmed closure of the defect with unobstructed pulmonary venous flow to the LA. There were no complications.
Conclusions: Transcatheter repair of SVD PAPVD is feasible using 3D printing techniques to select patients. This can be performed without complication avoiding cardiopulmonary bypass

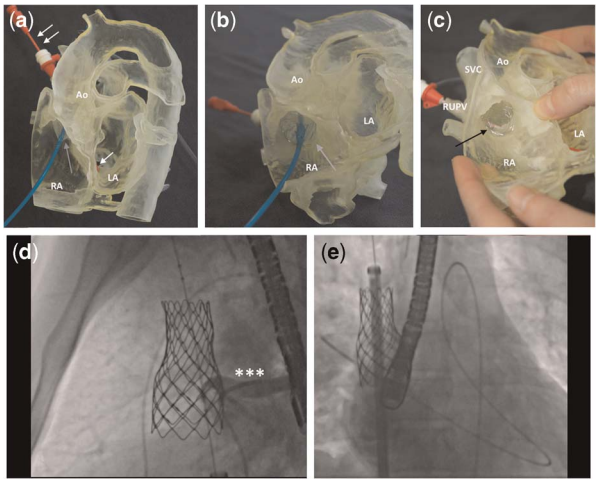

Figure 2.

P2437 - DUAL VENOUS CONNECTION OF PULMONARY VEINS WHY IS SO IMPORTANT TO DIFFERENTIATE IT FROM PARTIAL ANOMALOUS PULMONARY VENOUS RETURN (PAPVR) OUR CENTRE EXPERIENCE PHYSIOLOGICAL AND THERAPEUTICAL CONSIDERATIONS

Francisco González-Barlatay, Ana Méndez-Santos, Nagib Dahdah, Joaquim Miro
Sainte-Justine University Hospital, Pediatric Cardiology Service, Montreal-Canada

A dual venous connection of pulmonary veins with the left atrium and systemic veins has been described in the past. The prevalence has not been stablished, and at first sight seem to be a very rare condition, but this can be due to under-diagnosis. Some physiological particularities make this entity different to PAPVR. The connections with the left atrium predispose to have higher QP/QS than the expected for an isolated PAPVR, and in certain cases can also predispose to have a right to left shunt. We present our series of 5 patients which can represent the spectrum of clinical presentation, from a high QP/QS and right chambers dilatation to a right to left shunt whose initial sign was a cerebral abscess, and their therapeutic approach.
Conclusions: Dual venous connection of pulmonary veins is a particular entity that needs to be highly suspected to be diagnosed. A percutaneous approach is almost always the treatment of choice with very low morbidity and mortality.

P2442 - COR TRIATRIATUM DEXTER PERCUTANEOUS APPROACH TREATMENT IN A CHILD. FEASIBILITY OF THE TECHNIQUE AND RESULTS

Francisco González-Bartalay, Ana Méndez-santos, Marie-Josée Raboisson, Joaquim Miro
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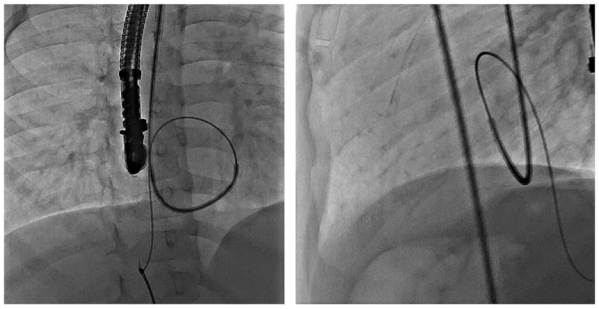
Remnants of the right sinus venosus valve can have several forms of presentation, from a fortuity echo finding to different degrees of right heart failure end cyanosis (Cor Triatriatum Dexter). The most frequent therapeutic approach of this very rare malformation is the surgical removal of the membrane. We report a new percutaneous technique performed in a toddler, which is the first case described in a pediatric patient.



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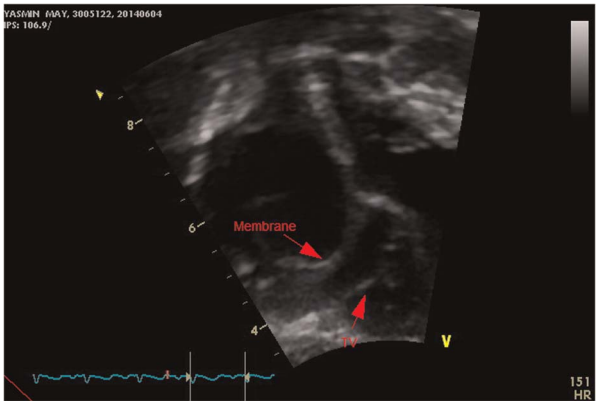

Figure 2.

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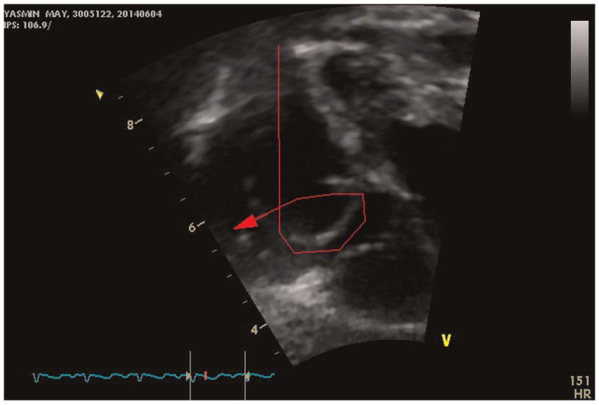

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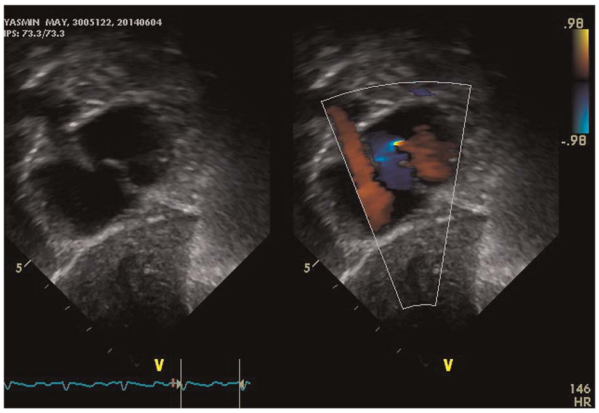

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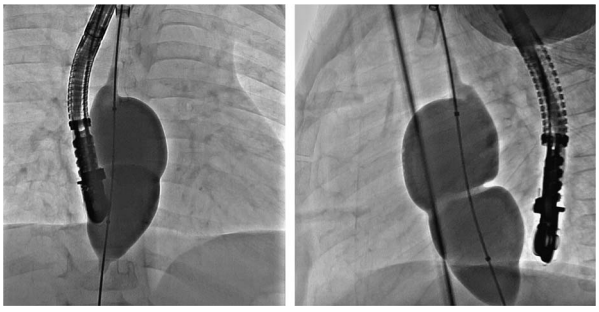

Figure 5.

P2446 - 10 YEARS SINGLE CENTER EXPERIENCE IN CARDIAC CATHETERIZATION IN HYPOPLASTIC LEFT HEART SYNDROME

Grace Van Leeuwen Bichara, Paulo Vasconcelos, Armando Mangione, Salvador Cristóvão
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Background/Hypothesis: Hypoplastic left heart syndrome (HLHS) patients are more likely to have catheter interventions as the prognosis of this condition has improved over the years. Various transcatheter procedures can be performed in order to access the hemodynamic and morphological status and to ameliorate the patient condition. We reviewed our institutional experience of transcatheter cardiac catheterization in HLHS patients.

Materials and Methods: Retrospective review of all HLHS patients who underwent cardiac catheterization from 2006 to 2016 in our institution.

Results: Eighty-six patients had a total of 159 transcatheter procedures (diagnostic or therapeutic). Eighteen procedures were performed after the first stage of palliation (post-norwood) (11%), at the mean age of 0,3 ± 0,1 years, 95 were after second-stage (post-Glenn) (60%) at the mean age of 2,2 ± 1,1 years and 46 procedures were after third-stage (post-Fontan) (29%) at the mean age of 6 ± 3,1 years. Therapeutic interventions were more commonly required in patients after second-stage (66%). Forty-six patients (53%) needed 71 therapeutic interventions. Interventions performed included collaterals arteries embolization = 44%, pulmonary arteries stents = 10%, aortoplasty using balloon or stent = 9%, venovenous channels embolization = 6%, balloon PA angioplasty = 6%, pulmonary vein angioplasty using balloon or stent = 2%, stent or balloon fenestration opening = 1%, occlusion of fenestration = 1%, venous system stent placement = 4%. Most of the procedures (75%) were performed after 2010 .

Conclusion: The optimal treatment of patients with HLHS is a process in evolution. Transcatheter procedures are commonly required and contribute for the success of the palliation. Most commonly we have performed diagnostic catheterizations and interventions after the second-stage of the treatment, preparing the patient for the Fontan procedure.

P2490 - OFF LABEL USE OF AMPLATZER DUCT OCCLUDER II; TRANSCATHETER RETROGRADE CLOSURE OF PERIMEMBRANOUS VSD IN CHILDREN SINGLE CENTER EXPERIENCE

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Objectives: To assess the safety and the efficacy of transcatheter closure of perimembranous ventricular septal defects (pmVSDs) by a retrograde transarterial approach using the Amplatzer Duct Occluder II (ADO II).

Background: Transcatheter closure of ventricular septal defects by antegrade method is well established technique, but require arteriovenous loop which add considerable time to the procedure, and the high incidence of developing atrioventricular block.

Material and Methods: This is a prospective study from a single center, from June 2015 to December 2016, 10 children were enrolled for Transesophageal echography (TEE) guided transcatheter retrograde closure of pmVSDs using ADO II device

Results: Successful closure was achieved in all patients. The mean age was 3.4 ± 1.58 years. The mean weight was 14.65 ± 3.93 kg. The mean VSD diameter measured with left ventricular angiography was 4.4 ± 0.52 mm at its exit. The mean VSD diameter measured with TEE was 4.4 ± 0.52 mm. All patients had aneurysmal tissue. Aortic rim was <3 mm in two patients. The mean Qp/Qs was 2.28:1 ± 0.20. The mean pulmonary artery pressure was 25 ± 3.33 mmHg. The mean fluoroscopic time was 15.9 ± 1.19 minutes. There were no procedure related complications, the follow up evaluation include ECG, TTE one day after the procedure, one month, 3, 6, and one year. Residual shunt was present in 20% of patients which decreased significantly to 10% 3 months later

Conclusion: Percutaneous retrograde device closure of pmVSD using ADO II device is a simple, safe and time saving technique in selected cases

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P2542 - A STUDY OF ANGLE FORMED BETWEEN PATENT DUCTUS ARTERIOSUS AND DESCENDING THORACIC AORTA IN BEATING HEART

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Pusan National University Yangsan Hospital, Pediatric Cardiology, Yangsan-Korea, South

Percutaneous transcatheter closure is an established method of treatment for patent ductus arteriosus (PDA). However, PDA device closure in infants or in patient with certain morphological types of PDA may be difficult due to protrusion of device into the descending aorta. However there has been few data about angles of PDA since 1951. In this study, we measured angles between PDA and descending aorta through angiography in beating heart. From Dec. 2008 to Nov. 2016, 190 patients who underwent percutaneous PDA occlusion was included in this study. Retrospectively, the angle of PDA was measured by 3 cardiologists between ampulla base of PDA and longitudinal axis of PDA through aortography. Patients were divided into 3 groups according to age (Group A: under 1 year, Group B: 1-6 years, Group C: more than 6 years). And also we stratified the patients by the PDA type according to Krichenko's classification. Of the 190 study patients, 134 patients were female. The median age was 7 years(range from 75 days to 60 years), and the average angle of PDA was $48.3 \pm 12.1°$. Compared with Mancini's results ($n = 24$; average angle of PDA $= 31.8 \pm 3.3°$), this study resulted in higher average angle of PDA($p < 0.001$). In Group A($n = 41$), average angle was $47.5 \pm 10.3°$. In Group B($n = 111$) and Group C ($n = 38$), average angle was $47.1 \pm 11.2°$ and $52.3 \pm 15.3°$ respectively. According to Krichenko's classification, 136 patients was included in type A("conical") and average angle was $48.5 \pm 11.6°$. In type C("tubular") and type E("elongated"), 21 and 32 patients were included and average angle was $49.7 \pm 18.2°$ and $46.6 \pm 9.2°$ respectively. In this study we measured the angle of PDA by using radiologic data from beating heart in bigger population. The average angle of PDA was 48.3°, which was higher than Mancini's results. We hope that our data will be useful to develop the new device for percutaneous PDA occlusion.

P2544 - ENDOVASCULAR STENT GRAFT REPAIR OF AORTIC DISSECTION TYPE B IN ADOLESCENTS

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National Heart Hospital, Pediatric Cardiology, Sofia-Bulgaria[1]; City Clinic, Cardiology, Sofia-Bulgaria[2]

Limited reports on the subject of aortic dissection in children and adolescents indicate that from 0.67% to 3.5% of aortic dissections occur in persons ≤21 years old. Rarely occurring in the pediatric population, aortic dissection is a condition with many predisposing factors: congenital cardiovascular disorders, connective tissue disorders, fibromuscular dysplasia, systemic hypertension and trauma. Surgical and endovascular procedures might cause the occurrence of aortic dissections. From September 2014 to December 2016 two adolescent patients underwent an endovascular stent-graft procedure for acute symptomatic type B dissection. Indication for endovascular repair is a complicated course of type B dissection, including visceral and/or peripheral ischemia, uncontrollable hypertension, and therapy-resistant pain. The first patient was a 15-year-old boy who at the age of 6 months underwent a surgery – PDA transection. He was hospitalized with accute chest pain. The computed tomography scan (CT) showed dissection of the descending aorta, Stanford type B. The mesenteric artery and left renal artery were involved. In first stage the mesenteric artery and abdominal aorta were stented. 20 days later, due to renal insufficiency, we implanted stent in the left renal artery. In the third stage the thoracic aorta was stented. The second patient was a 17-year-old boy with headache and high blood pressure. The patient's past medical, familial, and drug history was unremarkable. The CT showed aortic dissection type B, without abdominal arteries involved. Due to the presence of therapy resistant hypertension the patient underwent an endovascular procedure two weeks after the hospitalization. Three stents were implanted in the descending aorta. Aortic dissection is an important differential diagnosis in adolescents with acute chest pain, which is hard to diagnose. The early recognition and rapid treatment is lifesaving. Endovascular stent-graft repair of the thoracic aorta is an alternative to surgical repair.

P2554 - OUTCOMES OF AMPLATZER VASCULAR PLUG II (AVP II) DEVICE FOR OCCLUSION OF PDA – 2 CENTRE EXPERIENCE

Atul Kalantre[1], Mehnaz Atiq[2], Ram Ramaraj[1], Nilesh Oswal[1], Gordon Gladman[1], Ian Peart[1], Ram Dhannapuneni[3], Rafael Guerrero[3], Arjamand Shauq[1]
Alder Hey Childrens Hospital, Cardiology, Liverpool-United Kingdom[1]; Aga Khan University Hospital, Cardiology, Karachi-Pakistan[2]; Alder Hey Childrens Hospital, Cardiothoracic, Liverpool-United Kingdom[3]

Background: Patent ductus arteriosus (PDA) has significant variation in morphology. Different devices have been used but not all devices are suitable for all types of PDA. Few case series have been published on using Amplatzer Vascular Plug II (AVPII) for PDA. We have used AVP II mainly in long tubular type but also other shapes of PDA. We believe our series is the largest to report.
Objective: To evaluate use of AVPII device for PDA closure and incidence of complications.
Method: A retrospective study of all the data where AVPII was used for PDA closure.
Results: TABLE 1
50 patients underwent PDA closure by using AVPII from June 2014 to January 2017 in 2 different centres in children as small as 4.4 Kg. AVPII size ranged from 6mm to 14 mm. In all except one (neck approach) femoral venous approach was used to deploy the device. In all except one case, it was possible to close the PDA. All cases had complete closure except one with small residual shunt noted on the following day. One device embolised in the first 24 hours which was retrieved. There were no procedure-related complications. There was no increased gradient across pulmonary artery or aorta.
Conclusion: AVP II is a safe and effective device for all PDA closure especially Krichenko type C and type E.

Table 1. Results.

Demographics (N = 50)	Median	Range	Ductal Morphology[1]
Age (months)	15	3–144	A = 3/50 (6%)
Weight (Kg)	8.8	4.4–30	B = 1/50 (2%)
Narrowest ductal diameter (mm)	4.5	1.5–8	C = 21/50 (42%)
Length of duct (mm)	13	6–20	D = 1/50 (2%)
Diameter of aortic ampulla (mm)	8.5	4.5–14	E = 24/50 (48%)
Procedure time (min)	61	31–137	
Fluoroscopy time (min)	7.4	1.3–25	

[1] Krichenko classification of PDA morphology

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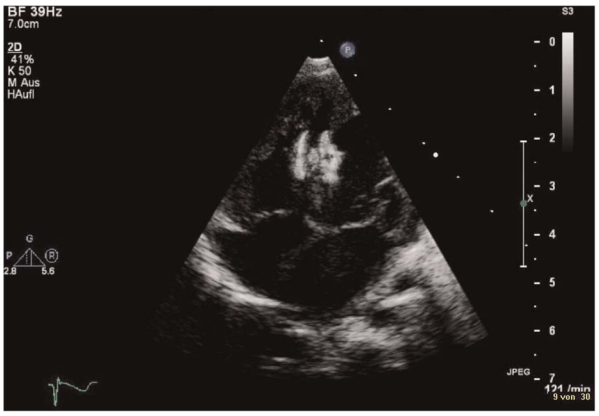
P2562 - OCCLUSION OF MUSCULAR VENTRICULAR SEPTAL DEFECTS IN SMALL INFANTS USE OF THE AMPLATZER DUCT OCCLUDER II

Stephanie Kleppe[1], Ulrike Herberg[1], Ralf Knies[1], Christine Bernhardt[1], Ingo Heinze[2], Oliver Dewald[3], Bahman Esmailzadeh[3], Johannes Breuer[1]
University Children's Hospital, Paediatric Cardiology, Bonn-Germany[1]; University Hospital, Anaesthesiology, Bonn-Germany[2]; University Hospital, Paediatric Cardiac Surgery, Bonn-Germany[3]

Background: Complete closure of haemodynamically relevant muscular ventricular septal defects (mVSD) can still be challenging by conventional surgery. Moreover, delivery sheaths for the available mVSD-occluders are relatively large when used for interventional procedures in infants.
Materials and Methods: Six patients aged three months to five years (weight 3-12 kg, median 7.39 kg) underwent interventional closure of a muscular VSD (5 apical, 3 midmuscular) with an AMPLATZER DUCT OCCLUDER II (ADOII). Two patients with multiple mVSDs received succesful implantation of a second ADOII in a separate session. All procedures were guided by transoesophageal (TOE) or transthoracic echocardiography (TTE). Vascular access was obtained via the jugular vein and the femoral artery via a 4 F sheath. The double-mushroom shape of the ADOII with its bigger rim and easy adapting to the rims of the septum allows choosing devices with exactly the same waist as the size of the mVSD measured by TTE or TOE. Via an arteriovenous loop the sheath was forwarded from the right jugular vein to the right ventricle (RV) and via the VSD into the left ventricle (LV). Then, the ADOII was advanced through this sheath and finally deployed.
Results: Seven procedures were completed successfully without facing major complications. A 3.5 months old girl showed transient significant bradyarrhythmias while forwarding the delivery catheter leading to abruption of the intervention. The mVSD was then closed with an ADOII using a hybrid approach. On follow-up of all patients no residual shunt across the devices was observed and no dislocation of any devices occurred.
Conclusion: The ADOII can be used safely even in small infants to close mVSDs not being suitable for a conventional surgical approach.



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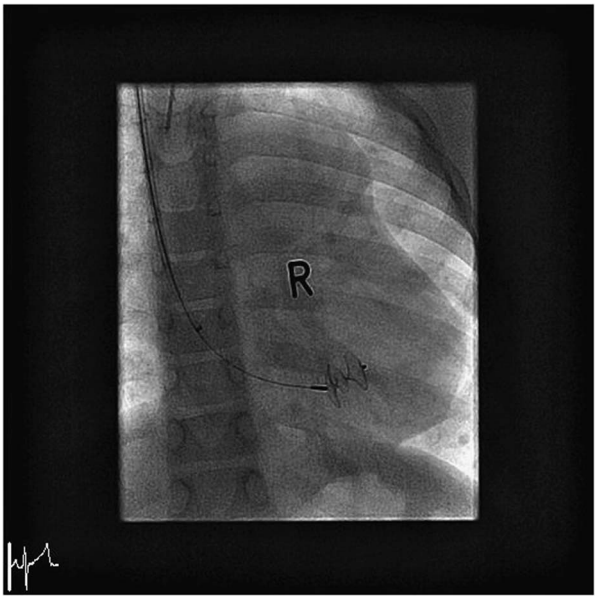
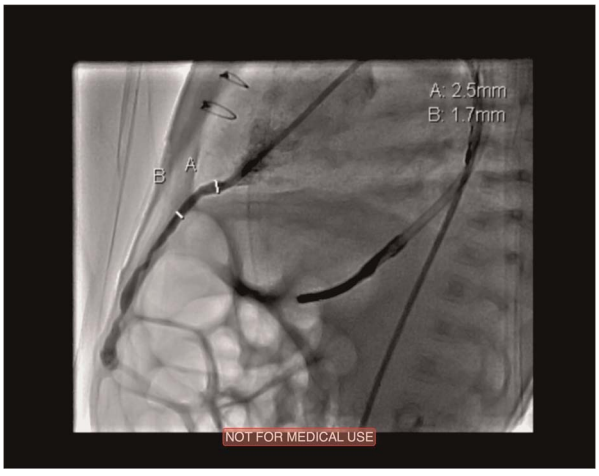


Figure 2.

P2567 - DEVICE CLOSURE OF A LEFT VENTRICULAR APICAL ANEURYSM A FIRST CASE REPORT

Mani Ram Krishna Singaravelu[1], Philip Roberts[1], David Murphy[1], Peter Grant[2]
Sydney Children's Hospital Network, Paediatric Cardiology, Westmead-Australia[1]; Sydney Children's Hospital Network, Cardiothoracic Surgery, Sydney-Australia[2]

Background: LV diverticula are outpouchings of the left ventricle with contractile myocardial tissue. They are classified into sub-diaphragmatic and supra-diaphragmatic diverticula. The sub-diaphragmatic type may be part of a Pentalogy of Cantrell. Surgical resection has remained the treatment of choice. We present a minimally invasive alternative to surgery in this condition.
Methods: This was a case report of a young infant who underwent per-cutaneous closure of a LV apical aneurysm. This is the frost reported intervention in literature.
Results: A neonate who was antenatally diagnosed to have a large VSD was evaluated postnatally at 15 days of life. Clinical examination suggested a moderately large shunt. In addition a pulsatile mass wa s seen in the epigastrium. Echocardiogram confirmed mesocardia with a large VSD. A small diverticulum was seen



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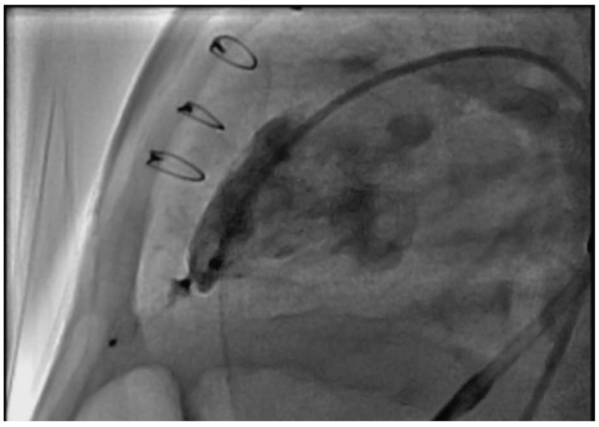
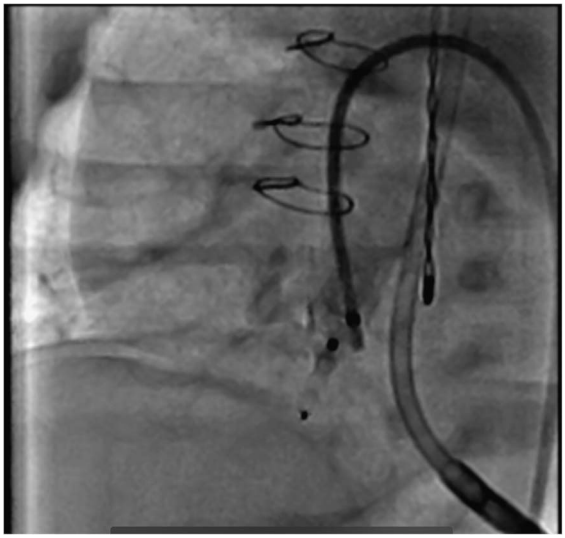


Figure 2.



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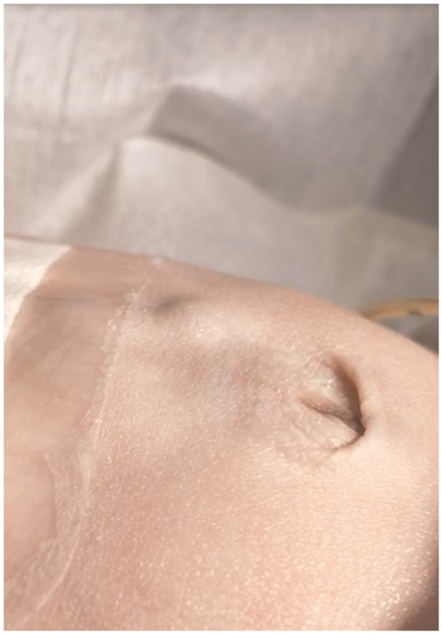

Figure 4.

arising from the LV apex and tracking below the diaphragm. The bay was initially palliate with a pulmonary artery band. Subsequently, cardiac catheterization revealed a restrictive 2, mm LV diverticulum apex. This was closed with a 3mm x 6 mm ADO Ii AS device. The pulsatile mass disappeared instantaneously and there were no residual flows.
Conclusion: Per-cutaneous occlusion with a device offers a safe and non-invasive alternative to management of infants with a LV apical diverticulum

P2574 - RETROGRADE TRANSCATHETER CLOSURE OF VENTRICULAR SEPTAL DEFECT USING AMPLATZER DUCT OCCLUDER II

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Jakarta Heart Centre, Pediatric Cardiology, Jakarta-Indonesia[1]; Cipto Mangunkusumo Hospital, Child Health Department, Jakarta-Indonesia[2]

Background: One of ventricular septal defect (VSD) transcatheter closure complications is total atrioventricular block (TAVB), occurring in 3% to 20% of cases. Butera et al. reported 6 of 104 subjects (5.7%) required pacemaker implantation. TAVB may occur immediately due to squeezing effects of disks or late due to device-induced inflammation and fibrous tissue formation. Amplatzer Duct Occluder II (ADO II, St. Jude Medical, St. Paul, Minnesota, USA) with nitinol wire and lack of patches inside the occluder was used in order to eliminate the risk of TAVB.
Materials and Methods: All datas of retrograde transcatheter closure of VSD using ADO II were collected from intervention database of 2 hospitals, Cipto Mangunkusumo Hospital and Jakarta Heart Centre Indonesia. We analysed data using IBM® SPSS® Statistics version 24. We selected weight of subjects at least 6 kg and VSD diameter less than 6 mm to be eligible for this study.
Results: Between January 2015 and December 2016, 35/54 patients underwent retrograde transcatheter closure of VSD using ADO II (19 male and 16 female). Their age ranged from 1 to 22 years (median 6.2 years) and weight ranged from 7.3 to 82 kg (median 21.4 kg). Median fluoroscopic and procedure time were 34 (10 to 89) min and 125.9 (50 to 190) min. Devices used (device diameter/length in mm) were 3/6 (n = 1), 4/5 (n = 1), 4/6 (n = 11), 5/6 (n = 10), and 6/6 (n = 12). Post-procedure of TAVB complication was not found. The defect was totally closed and there was no rhythm or conduction abnormality.
Conclusions: Retrograde transcatheter closure of VSD using ADO II can be performed safely and successfully. ADO II with gentle construction can be used in order to eliminate the risk of TAVB. Monitoring of rhythm still remain mandatory during long term follow-up.

P2575 - EFFICACY OF AMPLATZER DUCT OCCLUDER II IN NON DUCTAL POSITION

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Background: Amplatzer Duct Occluder II (ADO II) is actually designed for closing long ducts in infants. There are few reports of "off-label" use of ADO II in non-ductal positions.
Aim: To evaluate the feasibility and efficacy of ADO II in non ductal position by transcatheter method.
Material and Results: 102 cases of VSDs and 4 cases, one each of aorto right ventricular, aorto right atrial tunnel, infantile hepatic endothelioma, common iliac artery aneurysms and 2 coronary

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cameral fistulae were closed with ADO II of various sizes, formed the material for this prospective study. Age ranged from 28 days to 23 years (mean 9.1 years). 74 perimembraneous VSDs, 14 muscular VSDs,13 Gerbode defects, one midmuscular VSD with dextrocardia, were closed from retrograde approach by wire only technique. The shortest fluoroscopic time was 4.2 min, mean was 8.4 ± 4.1 min. Seven cases developed transient complete heart block and four cases had junctional rhythm, which resolved with oral steroids. Only one of them needed temporary pacing for 48 hours.
Discussion: ADO II has a very low profile, as there is no polyester material in it and can be easily delivered through a 4-5 F guiding catheter, needs very short fluoroscopic time as artero-venous (AV) loop is not needed in this retrograde approach. Cost is 1/3 the cost of regular ventricular septal occluder. However, it is not useful in VSDs measuring more than 6 mm and in those with insufficient aortic rim. Trackability of ADO II is excellent in rare, tortuous tunnels and fistulae.
Conclusions: ADO II is an excellent device in some non ductal positions like VSD, Gerbode defects, CAVF, ARVT. The procedure time and the cost is significantly lesser than regular devices. The success rate is very high and complication rate is very low.

P2582 - ASD DEVICE CLOSURE IS TRANSOESOPHAGEAL ECHOCARDIOGRAPHY MANDATORY

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Background: Transcatheter closure of Atrial Septal Defects (ASD) is generally considered to be best undertaken with trans esophageal echocardiography(TEE) to guide optimal placement of the device. However, in our experience, Transthoracic echocardiography (TTE) combined with specific fluoroscopic views provide sufficiently adequate imaging for undertaking safe ASD device closure procedures in children and thin individuals.
Materials and Methods: We report our experience of performing 185 device closures of ASD between April 2014 and January 2017 without TEE guidance. Age ranged from 2 years to 60 years and weight from 10 kg to 58 kg. TTE imaging was performed using subcostal sagittal & coronal views, four chamber & parasternal short axis views to determine the defect morphology, thickness or lack of it along defect margins and aneurysms of the septum. An allowance of 2-4mm more in device size was given for floppy or aneurysmal defect margins. Under deep conscious sedation or local anaesthesia, the device is deployed from the left or right upper pulmonary vein in Postero-Anterior (PA) view. Balloon sizing was undertaken in 12 patients when the chosen size of the device was found grossly inadequate. The seperation between the two discs astride the aortic torus in this view is used as a good guide to the suitability of device size. Device stability and all around disc seperation is assessed in LAO view with a gentle pull and push manoeuver. After TTE confirmation of device position, the release is effected in PA view.
Results: Immediate device embolisation occurred in 4 patients necessitating surgical retrieval and subsequent patch closure. There were no morbidities or mortality.
Conclusion: Apart from obese patients, it is feasible to safely undertake ASD device closure without TEE guidance. The avoidance of endotracheal intubation, general anesthesia and oesophageal echocardiography probe placement with its attendant risk is avoided.

P2589 - SIMULTANEOUS TRANSCATHETER TREATMENT OF COARCTATION OF AORTA COMBINED WITH PATENT DUCTUS ARTERIOSUS BY USING A COVERED STENT

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Background: Beyond infancy coexistence of coarctation of aorta (COA) and patent ductus arteriosus (PDA) occasionally occur. Age of the patient, anatomy of the coarctation, size and type of the ductus arteriosus all have importance while considering the treatment options for the patients with coexistent CoA and PDA. We will present our experience in transcatheter treatment of CAO combined with PDA.
Method: With the diagnosis of coarctation 198 patients underwent stent implantation between 2007-2016 in our institute. 12 of them had associated PDA. Hemodynamic assessment and pulmonary vascular reactivity testing for pulmonary hypertension performed in patients with severe pulmonary hypertension. Covered CP stents (CCP) were used in all.
Results: Median age was 15 (6.5-35 year). PDA type were type E in six, type C in four, Type A in one and type B in one. The median PDA diameter was 3.9 mm (2-9.9). The median peak to peak gradient was 43 (10-71) mmHg across the coarctated segment which was reduced to 0 (0-8) mmHg. The diameter of coarctated segment of aorta increased from a median 8.4 (2.6-10.8) mm to16 (9-24) mm after CCP implantation. Among four patients with severe pulmonary hypertension before the procedure, only in one of them it persisted (this patient had also large VSD). Fourteen covered stents used for 12 patients. After deployment of covered stent, four of them had residual PDA flow. Proximal part of the stents was flared with larger diameter semi compliant balloons. After flaring, there was no residual shunt across the PDA. CT angiography performed to all at the first year of intervention. None had recoarctation and no reintervention required during the median 20 months of follow-up.
Conclusion: By using a covered stent, COA combined with PDA can be treated safely and effectively in a single stage in older children, adolescents and adults.

P2596 - INITIAL EXPERIENCE WITH STENT IMPLANTATION IN THE RIGHT VENTRICLE OUTFLOW TRACT IN PATIENTS WITH TETRALOGY OF FALLOT

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Introduction: Tetralogy of Fallot (TOF) treatment is difficult in low birth weight newborns particularly when significantly ill or with unfavorable anatomy. Stent implantation in the right ventricle outflow tract (RVOT) is an option for them. We report our initial experience with RVOT stenting in patients with TOF.

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Methods: Retrospective and descriptive study conducted in 3 pediatric cardiovascular centers in Chile between 2012-15, including all TOF patients in whom a stent was implanted in the RVOT as first procedure. Clinical records, echocardiographic, interventional and surgical reports were reviewed for demographics and information of RVOT and pulmonary arteries (PA).
Results: RVOT stents were implanted in 12 newborns with TOF (9 female). Median age was 16 days (1–70). Mean gestational age was 34 weeks (28–40). Mean weight was 2.178 g (1.400–3.414). In 58% birth weight was below 2.500 grs. Oxygen saturation increased from 74,3% ± 7,3 (55–88) to 88,5% ± 4,0 (80–98%), ($p < 0,01$) after the procedure. No complications or mortality were related to interventions. Follow up was mean of 11 months (7–36). In 50% an additional procedure was needed. Mean PA diameters were at baseline and at the moment of surgery: right 2,3 ± 0,3 mm/5,3 ± 0,6 mm ($p < 0,05$) and left 2,2 ± 0,4 mm/ 4,0 ± 1,1 mm ($p < 0,05$). The surgical repair was performed median of 4 months (2-7) in 11 patients. No surgical mortality. The median postop stay was 7 days (6-32).
Conclusions: RVOT stenting is a safe and useful option for small patients with TOF and surgical risk factors or unfavorably anatomy. It increases the pulmonary blood flow, improving oxygen saturation and pulmonary arteries growth as bridge for surgical repair.

P2601 - SAFETY AND EFFICACY OF DUCTAL STENTING – SINGLE CENTRE EXPERIENCE

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Background: Ductal stenting (DS) is a less morbid alternative to surgical shunts in patients with duct-dependency.
Objective: To analyse patient profile, outcomes and complications of ductal stenting.
Methods: DS procedures in the past three years (2013–2016) in infants below 4 months were analyzed for procedural outcomes and follow up till their definitive procedure.
Results: Among 33 patients weighing 3.29 ± 1.26 kg (2.2-5.3 kg), 14 had PA in univentricular heart, 5 had TOF with PA, 1 had TOF, 2 had critical PS, 5 had PA and IVS and 1 was with large rhabdomyoma causing severe RV inflow obstruction required DS for pulmonary blood flows. 3 patients had retraining of regressed ventricle in transposition and 1 had revascularization of isolated left PA. 2 patients with non-confluent PAs needed bilateral DS. 1 patient with HLHS underwent DS as a part of Hybrid procedure. Procedural time was 91.51 ± 49.4 minutes (30-240 minutes) with saturations improving from 67 ± 11% to 88 ± 5%. Sirolimus/ Everolimus eluting stents were used in 14 patients. 5 patients needed additional stent to cover the entire ductal length. Procedural complications included arrhythmia in three, distal stent migration in three, 2 of which were stabilized with additional stent, low cardiac output state in six and retroperitoneal bleed in one. The mean ICU stay was 84.1 ± 141.8 hours(6 hours to 28 days). 3 patients had instent restenosis between 3 to 6 months. Ingrowth was not seen in drug eluting stents. Mortality occurred in 5 patients. Improvement of PA sizes were found in all. 6 patients had biventricular repair, while univentricular palliation was carried out in 5 patients. The mean duration of follow up for patients awaiting surgery was 444.53 ± 237.29 days, but still not hypoxic to warrant their next palliation.
Conclusion: DS is feasible, effective, safe procedure, which can act as a bridge to more definitive surgical intervention in the future.

P2614 - IMMEDIATE AND MID TERM RESULTS OF BALLOON AORTIC VALVULOPLASTY (BAV) FOR SEVERE CRITICAL AORTIC VALVE STENOSIS IN NEWBORNS. A SINGLE CENTER EXPERIENCE

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Assessment of immediate/mid-term BAV outcome in neonates with severe/critical AVS.
Material: 28 neonates with severe/critical AVS were referred to BAV. The group consisted nine premature infants (GA 34.7 ± 1.3 weeks, body mass 2.54 ± 0,4 kg). BAV was performed at the age: x = 12.8 ± 9 days, body mass: 3.14 ± 0.6 kg. Follow-up time was x-5.19 ± 2.9 years.
Methods: We analyzed LVEF (depending on values: ≤40% and >40%), initial and post-BAV transvalvular pressure-gradient (TVPG) and significant (≥3o) AV-regurgitation (AVR) incidence, re-BAV and/or surgical operation necessity. In follow-up, NYHA class of the patients, residual TVPG, LVEF and significant AVR incidence were analyzed.
Results: In 14 pts, LVEF was ≤40% (x = 28.3%) (GrI), while in the remaining 14 (GrII) - x = 63.3%. Post-BAV TVPG in GrI and GrII decreased (40.9 ± 23.3 mmHg to 29.2 ± 15.7 mmHg; 82.5 ± 22,2 mmHg to 40.7 ± 19.4 mmHg, respectively). Significant AVR was present in one GrII pts. Adverse events occurred in five pts: LV-wall perforation - two, AV-perforation - one, significant hemorrhage - one, and significant AVR 1 pts. Ten pts required re-interventions: two pts re-BAV, five - surgical valvuloplasty, two pts - hybrid-stage-one-procedure for HLHS, due to borderline LV and hypoplastic aortic annulus diameter, respectively, one child re-BAV at 3 months of age, surgical valvuloplasty when 3.5 years old and Ross-Konno procedure when aged 5 years. In follow-up, seven pts (25%) died (including 5/9 of preterm infants) at 19 days to 12 months post-initial BAV, including two post-hybrid-stage-one-procedure for HLHS. Fisher test showed significant factor of death to be prematurity ($p < 0.02$). In the groups, no significance was noted for body mass <2500 g. In late follow-up, all other pts were in NYHA I, residual TVPG in Doppler echo was x = 36.8 mmHg ± -13.6 mmHg, LVEF– x = 69.9% ±7%, three pts had AVR >2nd degree, and 10 pts ≤2nd degree.
Conclusions: Transcatheter treatment of severe/critical AVS is a life-saving procedure. Prematurity and probably borderline LV are poor prognostic factors in follow-up.

P2616 - RIGHT HEART CATHETERIZATION USING BASILIC VEIN APPROACH INITIAL EXPERIENCE

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Background and Objectives: Cardiac catherization is traditionally performed from groin using femoral vein and/or artery, which involves admission, limb immobilization and significant risk of bleeding and hematoma. Recent studies have shown safety of using arm vessels for cardiac catheterization. In this study we evaluated the feasibility of right heart catheterization using basilic vein.
Materials and Methods: We reviewed all patients undergoing right heart catheterization using basilic vein approach at our hospital

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from January 2013 to December 2016. Demographic data, indication, procedural and outcome data was analyzed using SPSS version 19.
Results: We reviewed all ~ 27 cases during the study period. Success rate in getting access was 100%. All patients were discharged home same day. Only two patients had small hematoma post-procedure. There was no clinical evidence of venous thrombosis at follow up in outpatient clinic.
Conclusion: Right heart catheterization using basilic vein approach is a promising technique in selected group of patients with shorter procedural time, early ambulation and more patient acceptance.

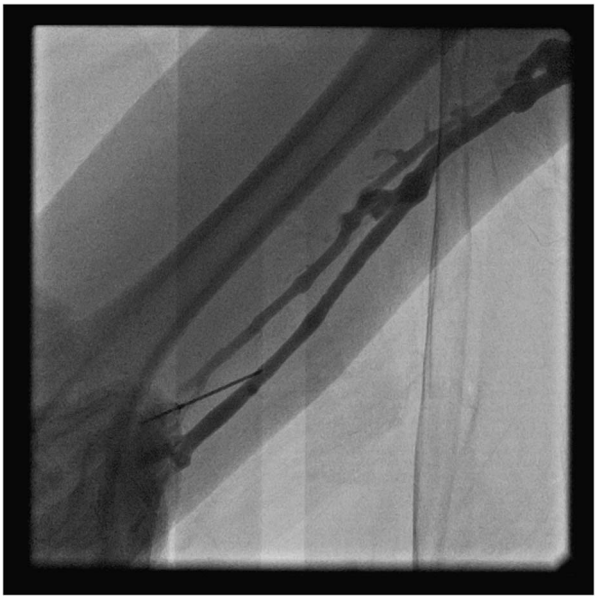

Figure.

P2624 - TRANSCATHETER CLOSURE OF FENESTRATIONS AFTER FONTAN TYPE OPERATIONS EARLY AND LONG TERM RESULTS

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Fontan type operations are the only surgical palliation with long-term positive hemodynamic effects in patients with single ventricle circulation. Conduit fenestration is a modification which shortens early postoperative adaptation. The presence of a fenester can have negative hemodynamic effect due to persistent right-to-left shunt with resulting hypoxia and paradoxical embolism. Transcatheter closure of the fenestrations improves exercise tolerance and quality of life.
Aim: Assesement of immediate and long-term results of transcatheter closure of the fenestration after Fontan surgery.
Material: Twenty children with single ventricle hemodynamics with completed stages of Fontan operation and subsequent transcatheter fenestration closure.
Methods: In all children the fenester was closed after comprehensive hemodynamic assessment in basal conditions and after temporary occlusion of fenestration. In 15 children the fenestration was closed by Amplatzer septal occlude, in 2 - by Cardioseal device, in 2 - using Figulla Flex II, and in 1 – by Amplatzer vascular plug. The patients were followed for a period of 6 months to 11 years.
Results: After fenestration occlusion oxygen saturation in the systemic circulation increased from $83{,}14 \pm 3{,}58$ to $92{,}58 \pm 3{,}20$ ($p = 0.0001$). Mean cavo-pulmonary pressure (mCPP) and pulmonary vascular resistance (PVR) showed no significant change: mCPP of 11 $28 \pm 1{,}70$ mmHg before occlusion increased up to $11{,}86 \pm 1{,}21$ mmHg ($p = 0.24$). PVR increased from $1{,}21 \pm 0{,}56$ to $1{,}51 \pm 0{,}51$ Wood units ($p = 0{,}15$). There were no procedural or late complications. In 6 of children, development of collaterals with a decrease in systemic oxygen saturation were found. In 5 out of these children, the collaterals were closed subsequently.
Conclusion: Transcatheter closure of fenestration in Fontan type surgery is effective and safe non-surgical treatment option. The accurate selection of patients is essential for good postprocedural results.

P2633 - FEASIBILITY AND SAFETY OF TRANSCATHETER CLOSURE OF ATRIAL SEPTAL DEFECT IN SMALL CHILDREN WEIGHING 10KG OR LESS

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Background: Transcatheter closure of atrial septal defect (ASD) has been accepted as a standard treatment. However, the efficacy and safety of transcatheter closure of ASD in very young and small children have not been clearly defined yet.
Methods: From 2005 to 2015, 124 patients (10.4% of total 1249 patients with device closure) weighing less than 10 kg were included in this study as device group. During same periods, 45 patients (12.8% of total 350 patients with surgical closure) less than 10 kg were included as surgery group. Baseline characteristics, hemodynamic features, comorbidities, success rate and complication rate were compared between two groups.
Results: Age (12.5 ± 5.4 vs 16.8 ± 6.1 months, $p < 0.001$) and weight (8.0 ± 1.8 vs 9.1 ± 1.0, $p < 0.001$) were lower in surgery group. However, hemodynamic features and anatomic details including rim deficiency (75.6% vs 73.4%, $p = 0.563$) and multiple defects (17.8% vs 27.4%, $p = 0.072$) were not different between two groups. Device closure was successfully performed in all children except 4 (3.2%) cases of mitral encroachment by atrial disc. Devices were retracted before detachment in these 4 patients and they underwent subsequent elective surgery. In both groups, there was no major complication. However, minor complication rate (1.6 vs 8.8%, $p = 0.04$) and hospital day (4.0 ± 0.3 vs 11.1 ± 5.4, $p < 0.001$) were significantly lower in device groups. In device group, two cases of transient arrhythmia were occurred. Catch-up growth was observed in both groups after ASD closure.
Conclusion: Transcatheter closure of secundum ASD is technically feasible, safe and effective even in small children weighing less than 10 kg. This study suggests device closure is a viable option in selected group of small children with ASD and there is no need to wait until they grow for transcatheter closure.

P2643 - TRICKS AND TECHNIQUES OF TRANSCATHETER CLOSURE OF CONAL SEPTAL VENTRICULAR SEPTAL DEFECT

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Introduction: Conal septum is the part of interventricular septum (IVS) which separates pulmonary valve from aortic valve. This

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muscular septum forms the supracristal part of IVS. Any ventricular septal defect (VSD) in this septum is known as conal septal VSD or outlet muscular VSD. These defects are located adjacent to the aortic and pulmonary valve and surgical closure is mainstay of treatment. Sometimes, this defect extends upto aortic valve and may produce aortic valve prolpase and aortic regurgitation (AR). Only a few case reports are available in literature about trascatheter closure of these defects. We are describing our experience of transcatheter closure of conal septal VSD.
Method: Total 5 patients were selected for transcatheter closure for conal septal VSD in Apollo Children's Hospital, Chennai in between April 2015 to March 2016. Mean age of the patients was 4 years (1-11 years). One of them had subaortic VSD with outlet muscular extension with mild AVP with no AR. Antegrade approach was used for 3 of them and VSD closure was done using duct occlude-I device. Retrograde technique was used for other 2 patients including the patient with subaortic VSD with outlet extension with AVP and the VSD was closed using duct occlude-II device. Mean follow up period was 9 months (3- 12 months)
Result: There were no major complications noted during the follow up period. None of them had any residual VSD. None of them had any left or right ventricular outflow obstruction or AR.
Conclusion: Conal septal VSD can be successfully closed by transcatheter technique. Proper case selection and proper assessment of the VSD is important for successful closure of this type of VSD. Longer term follow up is required to draw the final conclusion.

P2645 - TRANSCATHETER CLOSURE OF DOUBLY COMMITTED SUBARTERIAL VSD MIDTERM FOLLOW UP

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Transcather closure of ventricular septal defect (VSD) is feasible alternative of surgery. But only a few reports are available in literature on transcatheter closure of doubly committed VSD. VSD device closure are generally not attempted in these VSDs because the device can impinge on the aortic valve cusp which can cause worsening of AR. Fear of producing left and right ventricular obstruction is also high as the VSD is very close to aortic and pulmonary valve. In this article, we are summarizing our experience of transcatheter closure of doubly committed VSD.
Method: Total 8 patients were selected for transcatheter closure for doubly committed VSD in Apollo Children's Hospital, Chennai in between April 2014 to March 2016. Mean age of the patients was 10.4 years (8-13 years). All of them had mild AVP and 5 of them had trivial to mild AR. VSD closure was done by retrograde approach. It was done successfully in 7 patients. AVP appeared to be significant in LV angiogram in one child, and AR became severe after crossing the VSD regradely. That child was referred for surgery. Duct Occluder-II was used for all of them. Mean follow up period was 9.7 months (3- 24 months).
Result: There were no major complications noted during the follow up period. None of them had any residual VSD. None of them had any left or right ventricular outflow obstruction. 5 of them had trivial to mild AR before VSD device closure. None of them had any increase in severity of AR during follow up.
Conclusion: Doubly committed subarterial VSD can be closed by trancatheter technique and AVP with mild AR is not contraindication for device closure. Proper case selection is important. Longer term follow up is required to draw the final conclusion.

P2650 - BALLOON PULMONARY VALVOTOMY IN POST INTRACARDIAC REPAIR OF TOF WITH TRANSANNULAR PATCH WITH BICUSPID VALVE

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Introduction: Reconstruction of Right ventricular outflow tract (RVOT) using transanuular patch (TAP) is a standard technique of intracardiac repair (ICR) of Tetralogy of Fallot (TOF). One of the most popular techniques is TAP with bicuspid valve made of Polytetrafluoroethylene (PTFE). Significant RVOT obstruction is one of the commonest causes of reintervention after TOF surgery. In case of RVOT reconstruction with TAP with bicuspid valve, the PTFE valve can get crumbled or stuck resulting in RVOT obstruction. Surgical management in the form of resection and reconstruction is the standard approach, which is not free of risk and sometimes difficult because of previous surgery. Here, we are reporting our experience of balloon pulmonary valvotomy in post ICR of TOF with TAP with bicuspid valve using high pressure MAX-LD balloon. Patient 1: 2-year-old boy from Bangladesh, asymptomatic, came for routine follow up. He underwent successful ICR for TOF using TAP with bicuspid PTFE valve at the age of 1 year. Echocardiography showed severe RVOT obstruction due to stuck PTFE valve with moderate PR. Cardiac catheterisation showed systemic RV pressure. Pulonary valvotomy was tried first with a low pressure balloon, but it was unsuccessful despite of inflation at maximum pressure. Then pulmonary valvotomy was tried with high pressure Max-LD balloon and it was done successfully. Patient 2: 4-year-old girl from Tamilnadu presented with mild effort intolerance. She underwent surgery for TOF with TAP with bicuspid PTFE valve at 1 year of age. Echocardiography showed severe RVOT obstruction due to stuck PTFE valve with mild RV dysfunction. Diagnostic cardiac catheterisation showed near systemic RV pressure. Pulmonary valvotomy was done successfully with high pressure Max-LD balloon.
Conclusion: High pressure balloon can be successfully used for pulmonary valvotomy in case of severe RVOT obstruction in postoperative cases of TOF. Longer term followup is required to draw the final conclusion.

P2651 - OUTCOME OF NEONATAL CATHETER INTERVENTIONS A SINGLE CENTRE EXPERIENCE

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Aim: To study the indications and complications of all neonatal catheterization procedures done at a tertiary care centre.
Materials and Methods: The records of all neonates (age 1 – 30 days) who underwent catheterization procedures from July 2011 – to December 2016 at our centre were reviewed retrospectively. The indications, vascular access and outcomes were studied.
Results: Of the total 168 patients who underwent neonatal catheterization,74% were male. Median age and weight were 13.5 days (range 1 - 30) and 3.1 kg (range 1.4–5.5) respectively. Clinical presentation include cyanosis (56%), shock (8.7%), recurrent cyanotic spells (7.5%), failure (3.7%), sepsis (5%) and asymptomatic (13.7%). 15% of patients received prostaglandin infusion before procedure. PDA stenting was done in (39%), balloon pulmonary valvotomy (19.6%), balloon atrial septostomy (19.6%), balloon aortic valvotomy (10.7%), pulmonary valve

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perforation (4.7%), balloon coarctoplasty (5.3%), RVOT stenting (5.3%), coarctation stenting (2.3%), diagnostic catheterization study (1.7%), collateral closure (1.1%), PDA device closure and vertical vein stenting (0.5%). Femoral access was the commonest in all groups. Other unconventional accesses used were: carotid artery (n = 14), axillary artery (n = 22), right internal jugular vein (n = 2) and umbilical vein (n = 5). The complications encountered were transient bradycardia/hypotension (n = 18), transient pulse loss (n = 14), access related hematoma (n = 1), renal failure (n = 3), stent thrombosis(n = 2), stent embolization (n = 1), duct rupture (n = 1) and aortic dissection (n = 3). Overall mortality was 18 (10.7%), with maximum deaths among the PDA stenting group (12/67), followed by pulmonary valve perforation (2/8) balloon coarctoplasty (2/9), balloon aortic valvotomy (1/18) and balloon atrial septostomy (1/18). Among them, 55% were procedure related deaths and the rest were due to associated sepsis or aspiration

Conclusion: This study provides an insight into the common neonatal procedures and their complications at a tertiary centre. The complications are probably attributed to the poor pre-procedure hemodynamics as most of them presented with acidosis.

P2654 - TRANSCATHETER CLOSURE OF MUSCULAR VENTRICULAR SEPTAL DEFECT IN INFANTS WITH HEART FAILURE

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Introduction: Surgical closure is considered as the gold standard for infants with muscular ventricular septal defect (VSD) presenting with heart failure. We present a single center's experience on transcatheter closure of Muscular VSDs in symptomatic infants.

Methods: Medical records of infants with muscular VSDs.were analyzed between July 2011 to July 2016. There were 25 symptomatic infants with muscular VSDs.

Results: Of the 25 infants, eight were males. The median age at the time of procedure was 7 months (range 2-12 months) and median weight was 5 kg (range 3.3 -10 kg). All the infants presented with heart failure. The VSD was single in 15 and multiple in 10 Infants. The location of the defects were: Mid muscular in 8, anterior muscular in 7, posterior muscular in 5 and apical in 5 infants. Approach was retrograde in 5 cases and antegrade in 20cases. Antegrade route included foramen ovale approach (17). Femoral accesses was taken in 22 cases and 2 cases through internal jugular vein and 1 through carotid route. Procedure was successful in all but one infant. The device deployed were: Lifetech muscular device in 10 cases, Amplatzer muscular device in 9 cases and Amplatzer Duct Occluder II in 6 cases. Complications include transient bradycardia in one and vascular injury in one infant. There was one early mortality. Child with a large muscular VSD and Coarctation of aorta who presented with hemodynamic instability died 12 hours following VSD device closure and balloon coarctoplasty. There was one death 2 weeks following the procedure due to non-cardiac condition

Conclusion: Transcatheter closure of Muscular VSDs in Infants is a feasible in selected group of patients and is a safe alternative

P2668 - SYSTEMIC ARTERIOVENOUS MALFORMATIONS IN CHILDREN EXOTIC CONNECTIONS & INTERVENTIONAL STRATEGIES

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Background: Systemic arteriovenous malformations are by and large, uncommon. Most of them are acquired. Most cases of congenital arteriovenous malformations are confined to isolated case reports. We present a series of congenital systemic arteriovenous malformations seen in the pediatric cardiology department at a tertiary care centre with special emphasis on imaging, possible embryological basis and interventional treatment strategies.

Materials and Methods: This is a prospective study undertaken over the last one year. A total of 5 patients were seen. Age of presentation varied from 7 days to 13 years.

Results: Patient characteristics and arterial, venous connections with their management strategies are summarized in the table below.

Conclusions: Systemic arteriovenous malformations, being rare are difficult to diagnose in the pediatric age group. Presentations may range from being asymptomatic to features of cardiac failure. Diagnosis on echocardiography is challenging due to the unusual nature of sites of entry and exit with many being extracardiac. The embryological basis of these unusual communications is yet not well understood. Device closures of most of these may be accomplished successfully after precise identification of the nature of communication.

Table.

AGE	SEX	ARTERY	VENOUS DRAINAGE	SYMPTOMATIC	INTERVENTION
45 day	Female	Anomalous artery from Descending aorta	Portal vein	Abdominal distension, hepatosplenomegaly	4/6 ADO II
13 day	Male	Subclavian artery	Portal vein	Congestive heart failure	6/4 ADO II
5 y	Male	External carotid artery	Internal jugular vein	Pulsatile neck mass	5/4 ADO II
13 y	Female	Axillary artery	Axillary vein	Swelling over shoulder	Coil closure (3 coils)
1 week	Female	LPA	Left atrium	Murmur/asymptomatic	Follow up

P2670 - EARLY FAILING FONTAN ROLE OF CATHETER INTERVENTION

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Objective: Aim was to assess the role of catheter based interventions in stabilization of early failing Fontan operation.

Methods and Results: All records of patients (n = 51) who underwent Fontan Sugery from 2011-2016 at our hospital were analysed retrospectively. There were 13(25.4%) females and 38(74.5%) males. Mean age at time of surgery was 10 years (4 years- 33 years). There was one hospital death. The average hospital stay was 15 days (6 days to 35 days). The most important reason for prolonged hospital stay was persistent pleural drainage. Five patients who had prolonged pleural drainage despite all medical management underwent catheter based intervention. Two patients (3.9%) had antegrade flow in Fontan and underwent device closure of antegrade flow. Other two patients (3.9%) were subjected to creation of Fontan fenestration as their PA pressures were found to be higher on post operative catheterization study. Out of these two one patient also had a simultaneous balloon dilatation of distal pulmonary artery at the

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same sitting. One patient (1.9%) had anatomical narrowing at conduit and inferior vena cava (IVC) junction with large thrombus in inferior vena cava. In this child after thrombolysis, stent was deployed at IVC and conduit junction.
Results: All patients had significant reduction in pleural drainage over 24 hours and got discharged within 5 days of catheter intervention.
Conclusion: Prolonged pleural drainage is the most common problem of Fontan Surgery. Responsible factors should be meticulously worked out. Judious use of Catheter based interventions for some of these causes may save the failing Fontan.

P2675 - TRANSCATHETER CLOSURE OF VENTRICULAR SEPTAL DEFECTS USING NIT OCCLUD®LÊ VSD COIL

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Objective: Investigate the mid-term results of transcatheter ventricular septal defect (VSD) closure with the PFM Nit-Occlud® Lê VSD coil.
Background: In 1988, Lock described the first transcatheter VSD closure using a Rashkind double-umbrella. In 2002, Hajazi used an Amplatzer® device specific for perimembranous VSD (pVSD) closure with successfully results but a high rate of permanent atrioventricular block. For this reason, the use of the Nit-Occlud® Lê VSD coil started without any cases of permanent atrioventricular block in large case studies.
Methods and Materials: Thirty-three patients underwent an attempt of VSD closure with the PFM device between February/2014 and December/2016: 24 perimembranous, 1 Gerbode, 2 muscular and 6 postsurgical residual. Mean age: 7,8 years (4 months-16 years). Mean weight: 31,6 kg (5,8-79). The mean diameter of the VSD was 5 mm (3,5-9) and mean Qp/Qs was 1,65 (1,2-2,8). The Nit-Occlud® Lê VSD coil is made up by flexible nitinol spirals as a left-sided-cone. An intraprocedural transesophageal echocardiography was performed. The follow-up protocol included echocardiography and electrocardiogram on day 1 and 7 after VSD closure and during 1°,3°,6° and 12° month.
Results: The device was successfully placed in 21 of the 24 pVSD, except for 2 cases of Laubry's syndrome and one large defect, and was successfully implanted in all the other type of VSDs. Complete closure rate in pVSD was: 15 immediately closure (15/21 = 71,45%) and 20 after 1 year (20/21 = 95,3%). In the other type of defects there were a complete immediately closure in all cases but one with mild residual leak. As complications, there were 3 cases of transient asymptomatic nodal rhythm.
Conclusions: The Nit-Occlud® Lê VSD coil device can be used successfully for perimembranous, muscular and postsurgical residual VSDs closure. As an important advantage, there was no permanent atrioventricular block. Despite the initial satisfactory results, larger series are necessary.

P2677 - BALLOON COARCTOPLASTY IN INFANTS WITH INTERMEDIATE TERM OUTCOME IT IS WORTH PURSUING

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Background: Coarctation of aorta carries significant morbidity and mortality in infants and neonates. Balloon coarctoplasty is not recommended as a procedure of choice due to reports of significant restenosis. We have reviewed the data from December 2009 to August 2015 of neonates and infants undergoing balloon coarctoplasty
Material and Methods: All babies with significant coarctation of aorta requiring intervention below one year of age were included. Gradients of more than 20 mmHg with normal LV function or any gradient with anatomic narrowing in presence of LV dysfunction were considered for intervention.
Results: 33 children underwent balloon coarctoplasty. 8 were neonates. M:F::3.8:1(26:7). Median age was 3 months (13days -12mon). LV dysfunction was present in 21(65%) babies.. 28(84%) babies had significant drop in pressure gradients to less than 30%. 3 (9%) babies had drop in pressure gradients between 30% - 50%. Average pressure gradients reduced from 41 + 30 mmHg to 6.7 mmHg. 2(6%) babies had gradients of >50%. There were no procedure death or dissection.
Follow up: 31(94%) babies had immediate success. Two babies died due to non cadiac causes. LV dysfunction resolved in 3 months. 3 babies were lost to follow up. Three babies had recoarctation at 3 - 4 months follow up(10.7%- 3/28). One neonate underwent repeat balloon coarctoplasty and two were referred for surgery. Mean follow up was 6mon(1mon -60 mon). Among 8 neonates seven had successful procedure(88%). One baby is lost to follow up. Among six, three required reintervention (50%), 3 are symptom free.
Conclusion: Residual gradients <30% predicts good long term outcome and >30% suggests need for reintervention. Residual gradients of more than 50% require surgical correction. Immediate success rate is 94%. Recoarctation is 10% in babies above 1 month and 50% in neonates. Recoarctation is unlikely after 1year. LV dysfunction resolves within 3 months.

P2680 - MEDIUM TERM FOLLOW UP OF TRANSCATHETER CLOSURE OF PATENT DUCTUS ARTERIOSUS USING AMPLATZER DUCT OCCLUDER II

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Background: Limited follow-up data of Amplatzer duct occluder II (ADOII) is available.
Objective: To see the efficacy and medium term results of transcatheter closure of PDA using ADOII.
Materials and Methods: This is a single centre follow-up study of patients underwent PDA closure using ADOII between 2010 and 2013. All patients were followed up and analyzed various parameters by transthoracic echocardiography and CT angiogram in selected cases. The angiograms of the procedure were reviewed and various parameters were analyzed retrospectively to see the cause for left pulmonary artery stenosis.
Results: Total 52 (female n = 31) patients underwent PDA device closure using ADO II. The median age and weight were 24 months (range 7-120) and 10.5 kg (range 5.5-25) respectively. Mean ductus arteriosus diameter was 3.6 2.8 mm. Analysis of angiogram showed type A in 36, type B in 1, type C 8, type D 4 and type E 3 cases. Size of the device used were: 3 × 4:2, 3 × 6:3, 4 × 4:9, 4 × 6:4, 5 × 4:9, 5 × 6:5, 6 × 4:12, and 6 × 6:8. The immediate occlusion rate was seen in 88%, within 24 hours 96% and 100% closure at 6weeks. There

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were no procedure related deaths. Vascular injury was seen in 1, device embolization in 1, device repositioning in1 and device was removed in 1. The median follow up was 4.8 years. Three cases developed significant left pulmonary artery stenosis confirmed by CT angiogram and cardiac catheterization (1). The analysis of various parameters showed young age, large device and distance between the device to 50% of left pulmonary artery diameter are the predictors of LPA stenosis. One child developed mild coarctation of aorta and became normal during follow-up.
Conclusion: Follow-up assessment is mandatory to see the left pulmonary stenosis in patients after transcatheter closure of Ductus arteriosus using ADOII.

P2685 - TRANSCATHETER CLOSURE OF VENTRICULAR SEPTAL DEFECT IN A CHILD WITH ANATOMICALLY CORRECTED MALPOSITION OF GREAT ARTERIES

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Background: Anatomically corrected malposition of great arteries (ACMGA) is an extremely rare form of congenital heart disease characterized by abnormal spatial relation of the great arteries maintaining atrioventricular and ventriculoarterial concordance. Structural defects like ventricular septal defects, atrial septal defects, subaortic and subpulmonary obstructions, juxtaposed atrial appendages are commonly associated with this condition. Till date there are no reports of transcatheter closure of ventricular septal defects in this subset of patients. We present a child with ACMGA who underwent successful transcatheter closure of VSD.
Materials and Methods: SK, a 8 year old girl, had dyspnoea on exertion with stunted growth. Echocardiography revealed a 5 mm upper muscular ventricular septal defect with significant left to right shunt. Angiograms at anteroposterior and 300 right anterior oblique views further corroborated the echocardiographic findings. A soft device in the form of Amplatzer Duct Occluder II (ADO II) was chosen to minimize the risk of possible complete heart block and to avoid subaortic obstruction following placement of a bulky device as prominent subaortic conus is associated. The defect in our patient was splayed on the right ventricular side, limiting the opportunity to close it from the arterial side. After obtaining an arteriovenous loop, a 6 × 4 ADO II was thus deployed from the venous side to successfully close the defect.
Results: The defect was totally occluded with no residual flow across it. Alienation of the device to the interventricular septum was perfect and the patient was in sinus rhythm with no obstruction to any outflow tracts at 6 months follow up.
Conclusions: Vivid understanding of the anatomy, particularly the alignment of the ventricles and the outflow tracts are essential, especially to profile the VSD angiographically. Thorough knowledge of the conduction system in ACMGA is a must before attempting any intervention in such patients.

P2689 - STENTING OF THE DUCTUS ARTERIOSUS IN NEWBORNS WITH DUCTAL DEPENDENT PULMONARY CIRCULATION 7 YEARS OF A SINGLE CENTER EXPERIENCE

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Objetive: describe our experience in ductal stenting on patients with ductal-dependent pulmonary circulation.
Background: Since the early 90 s, ductal stenting has been proposed to maintain ductus patency as an alternative to surgical shunts to ensure a stable pulmonary blood flow. In the last decade, the technique has become a viable alternative as a result of device's and technical improvements.
Methods: We retrospectively analyze 27 patients who underwent cardiac catheterization for ductal stenting in our center from March 2009 to September 2016.
Results: The procedure was successfully completed in 17 patients (65,3%, Table 1). One patient died after anesthetic induction before

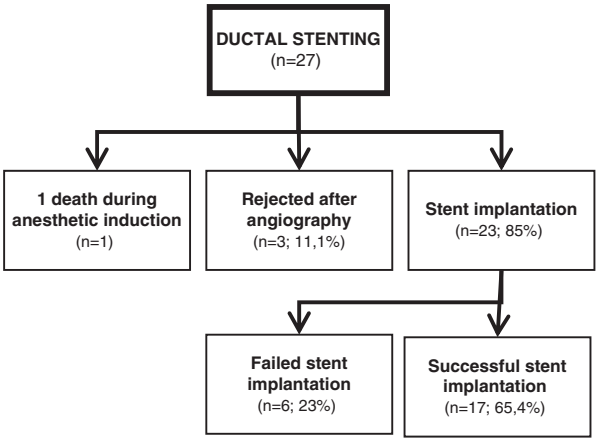

Figure 1.
Flow chart of the total patients

Table 1. Demographic data of patients who underwent successful stent implantation

Demographic data	Value
Age (days), mean (range)	12 (1 – 30)
Weight (kg), mean (range)	3 (2 – 4,2)
Procedure time (minutes), mean (range)	155 (65 – 290)
Diagnosis (n):	
- Biventricular physiology:	12 (70,59%)
Tetralogy Fallot, pulmonary stenosis	4
Double outlet right ventricle, pulmonary stenosis	1
Pulmonary atresia with intact ventricular septum	2
Pulmonary atresia with ventricular septal defect	2
Critical pulmonary stenosis	1
TGA, ventricular septal defect, pulmonary stenosis	2
- Univentricular physiology:	5 (29,41%)
Pulmonary atresia with intact ventricular septum	1
Ebstein´s anomaly	2
Tricuspid atresia	1
Complex congenital heart disease*	1
Ductus arteriosus (mm), mean (range):	
- Aortic side	4,08 (2,4 – 6,5)
- Pulmonary artery side	1,9 (0.8 – 3,8)
Stent (mm), mean (range):	
- Diameter	4 (4 – 4,5)
- Length	13 (8 – 18)
Stent implantation:	
- Via axillary artery (3 or 4Fr sheath)	7 (41,2%)
- Via femoral artery (4Fr sheath)	7 (41,2%)
- Via femoral vein (5Fr sheath)	3 (17,6%)

TGA: transposition great arteries. * Complex congenital heart disease: single right ventricle, common atrioventricular valve, malposition of the great arteries and pulmonary stenosis.

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starting the procedure because of haemodynamic deterioration. There were no other procedure-related deaths. In 3 patients stent implantation were not attempted because of an adverse anatomy at the angiography: bilateral pulmonary artery stenosis (n = 1) and ductal tortuosity (n = 2). In 6 patients, the stent could not be implanted or embolized. These 9 patients were referred for surgery. The freedom from reintervention (surgical shunt n = 1/re-catheterization n = 9) was 126,1 days (2-381). Average time from stent implantation to the first cardiac surgery was 294 days. Four patients required early re-catheterization (<3 months) due to: ductal stenosis (n = 2), incomplete ductal stenting on the aortic side (n = 1), and pulmonary over-circulation (n = 1). Mayor complications occurred in four patients (3 stent embolization and 1 femoral dissection) and minor complications in nine (3 transient arrhythmias, 5 vessel entry thrombosis, 1 intimal left subclavian artery dissection). The number of referred patients for ductal stenting has tripled in seven years. In 2009 half of the procedures failed, while since 2014 all of the ductal stents have been successfully implanted.
Conclusions: Ductal stenting is reasonable, feasible and safe as an alternative to surgical shunt in newborns with ductal-dependent pulmonary circulation. The results are satisfactory after the learning curve.

Table.

	Duration of symptoms	Hepatic Vein cannulated	Succesful	Balloon/ Stent	Follow up
1. 4 yr/F	1 month	Right hepatic vein	Successful	Balloon with stent insertion	Four year asymptomatic
2. 6 yr/M	2 months	Left hepatic vein	Immediate success	Balloon	Improved in 10 days, lost to follow up
3. 8 mon/M	3 days	Right hepatic vein	Successful	Balloon	Died on 6th day with variceal bleed
4.9 y/M	1 month	Left hepatic vein	Successful	Balloon	Two years follow up, asymptomatic
5. 2 yrs/F	5 months	Unsuccessful	Unsuccesful		Died due to variceal bleed
6. 1.5 yrs /M	1 month	Unsuccessful	Unsuccessful		Died within a month
7. 3 yrs/M	4 months	Unsuccessful	Unsuccesful		Died within one week with encephalopathy and sepsis
8. 16 y/M	4 mon	Right hepatic vein/IVC obstruction	Successful	Stent to IVC	Follow up -2 yr Asymptomatic
9. 14 y/F	1 week	Unsuccessful	Sent for surgery		Died due to coagulopathy
10. 9m/F	2 weeks	Right hepatic vein/IVC obstruction	Successful	Balloon with stenting after 2 months	Follow up 12 months

P2692 - INTERVENTIONAL THERAPY IN PEDIATRIC BUDD CHIARI SYNDROME – GRATIFYING OUTCOMES

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Background: Budd- Chiari syndrome (BDCS) is an eponym for hepatic venous out-flow obstruction It leads portal hypertension, hepatic dysfunction and cirrhosis. Establishing patency of the hepatic veins is the most desirable therapy. The outcome for Budd Chiari syndrome is dismal without treatment. Surgical portosystemic shunting carries high morbidity is of palliative nature.
Materials and Methods: Data collected retrospectively and prospectively from January 2012 - December 2016 from hospital records
Results: 10 children presented to our hospital with Budd- Chiari syndrome. Interventional therapy was attempted in all 10 children but was successful in 6 of them. Initial balloon dilatation was done. Stenting was necessary when there was a gradient of more than 2 mmHg and narrowing on angiogram after balloon dilatation. Stent implantation was necessary in 3 children. All children who underwent intervention became completely asymptomatic within 10 days. Longest duration of follow up is 4 years. Among six children with successful intervention four are on follow up and at the end of four years are doing well with normal portal pressures and liver function. The ascites has completely disappeared. Their growth has normalized. All are on anticoagulation. One child died within a week of intervention. It had acute Budd chiari and developed severe variceal bleeding. One child is lost to follow up. One child developed recurrence and required stenting at 3months on follow up. All four children where intervention was not possible were referred for surgery but unfortunately died while awaiting surgery or in immediate postop period.
Conclusion: Hepatic vein angioplasty is the most physiological therapy for Budd –Chiari syndrome. It should be attempted for every child before embarking on surgical options. Long standing obstructions are difficult to intervene. When successful the results are gratifying and intermediate term follow up shows event free survival.

P2727 - UTILIZATION OF THE EDWARDS SAPIEN 3 VALVE IN THE NON AORTIC POSITION IN CONGENITTAL HEART DISEASE

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Background: Transcatheter pulmonary valve replacement with the MELODY valve has become an important adjunct in the treatment of children and adults wiht failing right ventricular outflow tract conduits. Recently, the MELODY valve has also been succesfully implanted in the tricuspid, mitral and aortic positions, typically within failing bioprosthetic valves. The Edwards SAPIEN 3 has been developed for treatment of calcified aortic valves in elderly patients at high surgical risk. There is limited data on the use of this new third generation valve in other positions.
Materials and Methods: We implanted three Edwards SAPIEN 3 valves transfemorally in the pulmonary position in one adolescent and two young adults with repaired tetralogy of Fallot by for severe pulmonary insufficiency without complications. All had received a transannular patch for reconstruction of the right ventricular outflow tract at previous surgery. The fourth patient had single ventricular physiology and he had undergone a Damus-Kaye-Stansel procedure, partiaaly reconstructed with a Dacron prothesis. The pulmonary portion of the Damus-Kaye-Stansel was severly insufficient in this symptomatic young adult. An Edwarsd. SAPIEN 3 valve could be implanted via the femoraly artery after pre-stenting the pulmonary portion of the Damus-Kaye-Stansel anastomosis.
Results: All four Edwards-SAPIEN 3 valves were not or not more than mildy stenotic or regurgitant at 6, 6, 12 and 15 months, respectively. No stent fractures or endocarditis occured.
Conclusions: The Edwards SAPIEN 3 valve may be an additional alternative to the Melody valve and previous versions of the Edwards Sapien valves through its combination of a smaller sheath size and larger diameter valves.

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P2734 - TRANSCATHETER OCCLUSION OF PERIMEMBRANOUS VENTRICULAR SEPTAL DEFECTS BY AN OCCLUTECH DUCT OCCLUDER DEVICE THE FIRST REPORTED CHILDREN

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Background: Ventricular septal defects (VSDs) are the most common congenital cardiac malformation. Standard treatment for perimembranous VSD (pmVSD) is open surgery. Transcatheter VSD closure avoids extracorporeal circulatory support and the surgical approach and has benefit of faster recovery, but both methods carry potential risk of atrioventricular block (AVB). Valvular regurgitation was another major consideration in transcatheter closure of pmVSD. We report, firstly, the use of the percutaneous closure of pmVSDs with Occlutech®duct occluder device, as an "off-label" in the five children (Fig. 1).
Materials and Methods: We reviewed, retrospectively, 5 patients with perimembranous VSD who underwent transcatheter VSD closure with device during the last year. The femoral approach was used in all patients. A long sheath was introduced into femoral vein over the arteriovenous circuit wire and advanced through VSD to aorta(Fig 2).
Results: All of patients were successfully treated by transcatheter intervention. Range of pmVSD size was 3.4- 4.5 mm, two of them whose are multipl pmVSD with originating from aneurysmatic tissue. There were used to close defects Occlutech® duct occluder with having a proximal diameters of 6-7 mm(D3), and distal diameters of 4-5 mm(D2) (Fig. 1). There were aortic valve with mild regurgitation in one patient, and trivial residual shunt with multipl pmVSD in another patient. There was no tricuspid regurgitation or AVB after procedure in any patients. Demographic and diagnostic data are summarized in Table 1.
Conclusion: The number of devices used for percutaneous closure of pmVSDs is very limited. The Occlutech® duct occluder, because of it has no right ventricular disc, can not hold aneurysmatic tissues created by tricuspid valve. The risk of developing complete AVB and tricuspid valve failure is low because of it's flexibility and absence of right ventricular disc. However these possible benefits should be supported in larger cohort studies.

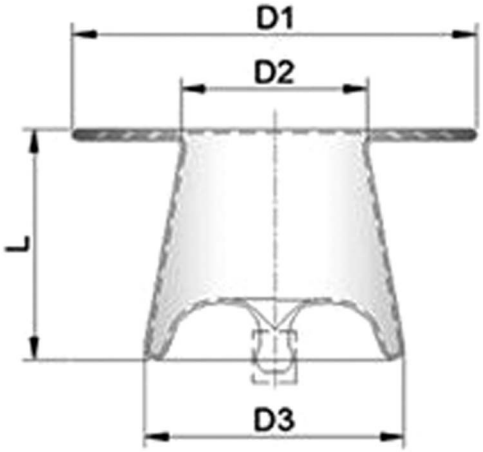

Figure 1.
Occlutech duct occluder

Table 1. The features of cases

	Case 1	Case 2	Case 3	Case 4	Case 5
Gender	Female	Male	Female	Female	Male
Age(years)	17	14	6	6	15
Weight(kg)	63	54	19	24	64
VSDsize (echo/mm)	3.6/1.5	4	4	3	4/3
VSDsize (angio/mm)	3.4/1.5	4.2	4.4	3.4	4.5/2,5
Fluoroscopic time(min)	56	48	35	84	50
Procedure time(min)	112	134	108	172	146
Qp/Qs	2.8	1.8	2	1.7	1.7
Device size (D2/mm)	4	5	4	4	5
Hospital stay (day)	5	5	5	5	5
Follow up (mo)	8	4	3	3	3
VSD types	Perimembranous/ multipl Aneurysmatic	Perimembranous	Perimembranous	Perimembranous	Perimembranous/ multipl Aneurysmatic
Tricuspid/aortic insufficiency	No	No	No	Aortic regurgitation (mild)	No
Residual shunt	No	No	No	No	Trivial
ECG abnormality	No	No	No	No	No

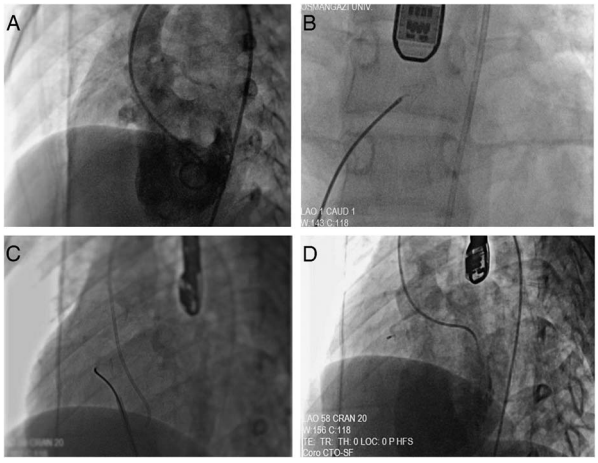

Figure 2.
(a) Left ventriculogram left to right shunt with VSD; (b,c) position of the device before being released; (d) after closure of the pmVSD with Occlutech duct occluder (5/7 mm), no residual shunt was seen.

P2745 - PERCUTANEOUS CLOSURE OF RIGHT PULMONARY ARTERY TO LEFT ATRIAL FISTULA

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Introduction: Right pulmonary artery (RPA) to left atrial(LA) fistula (PALAF) is a very rare cardiovascular anomaly that results in central cyanosis. We report the multiple brain abscesses associated with PALAF and successful closure using Amplatzer Muscular VSD occluder device in a child. Case Report A 8 years old boy was admitted with cough and cyanosis. Physical examination was performed that cyanosis and clubbing fingers were detected. Echocardiography showed elongated left atrium and a communication between RPA and LA. Agitated saline injection showed early appearance of contrast bubbles in the LA within two beats. When the patient complained of blurred vision, the story was

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deepened and the patient was informed that he had complained of headache for 20 days. Brain tomography was performed and multiple brain abscesses was detected (figure 1). Abscess drainage was performed and brain abscess treatment was completed with antibiotic therapy. After the brain abscess treatment cardiac catheterization was performed. The narrowest part of fistula was 10 mm in balloon sizing (figure 2). A 12 mm Amplatzer Muskular VSD occluder device was deployed at the narrowest site and the fistula was closed successfully (figure 3). Control angiography demonstrated complete occlusion of fistula. After the procedure, arterial oxygen saturation in room air immediately increased from 74% to 95%.

Discussion: PALAF bypasses the capillary bed; the lung loses its filtering function, thus bacteria to pass directly into the systemic circulation, resulting in cerebral abscess. Untreated cases can cause high mortality complications such as systemic thromboembolism, infective endocarditis, brain abscess, aneurysm rupture. Early diagnosis and treatment are very important. In select cases, transcatheter closure appears to be a safe and effective alternative to surgical treatment.

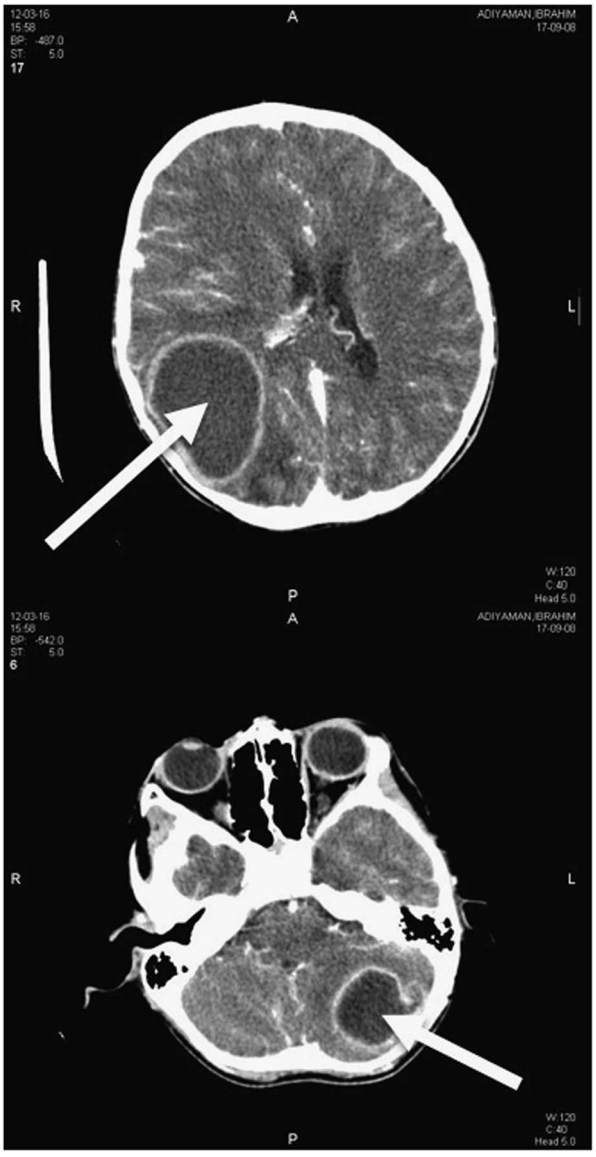

Figure 1.

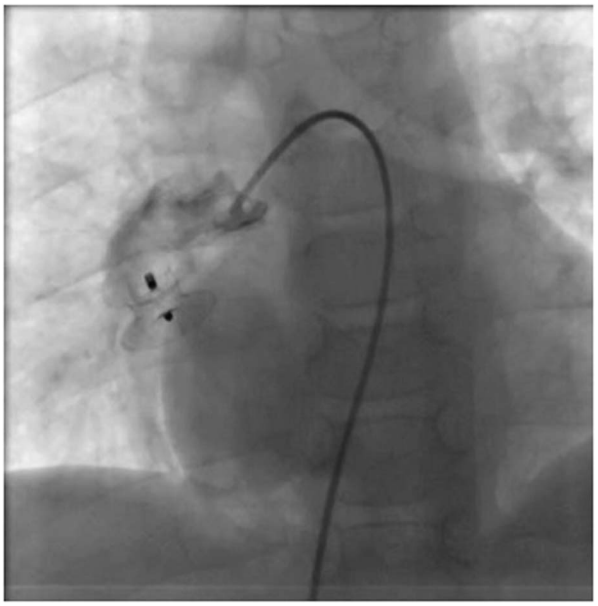

Figure 2.

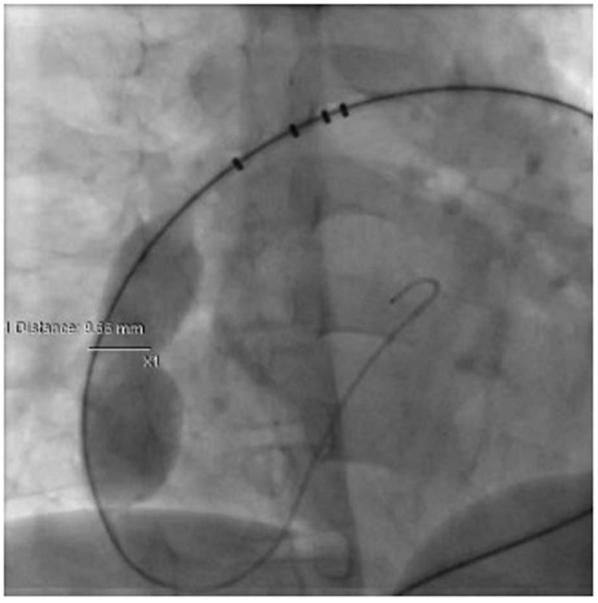

Figure 3.

P2746 - THE USE OF STEERABLE GUIDE CATHETER DURING TRANSCATHETER CLOSURE OF ATRIAL SEPTAL DEFECT IN A CHILD WITH INTERRUPTED INFERIOR VENA CAVA CASE REPORT

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Background: Percutaneous closure of trancatheter secundum atrial septal defect (ASD) is the first option for defects with adequate rims. Femoral vein is the conventional access route for the

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transcatheter procedure but congenitally interrupted inferior vena cava may make this route impassable. Our report is the first Turkish study that describes the closure of an ASD with an Agilis steerable guide catheter using the transjugular access.
Case: The patient, 12-year-old girl, was admitted to our department with the complaint of easy fatigability. A transthoracic echocardiogram (TTE) revealed ASD measuring 12-mm in diameter. Infrahepatic interruption of the inferior vena cava with azygous continuation is seen in the TTE and contrast study (Fig. 1). Hence, a 6-Fr sheat was placed into the right internal jugular vein. Attemps at deployment of the device ineffectived while the device removed through the delivery sheat because the exchange guidewire was removed the from inferor pulmonary vein. The 13-mm Amplatzer septal occluder device was loaded and delivered to the left atrium through the 8,5-Fr Agilis steerable curved catheter. Then, the left and right atrial discs were fully opened and the device deployed under fluoroscopy with continuous transesophageal visualization (Fig. 1). A transesophageal echocardiography showed the elimination of any shunt. The patient was discharged home uneventfully at the next day.
Conclusion: Agilis, steerable guide and adjustable curved catheter, used with the transjugular approach proves guidewire advancement into the pulmonary vein and correct engagement of the deployment device. It may be of use in patients with impaired or lost inferior vena cava continuation, such as in interrupted IVC.

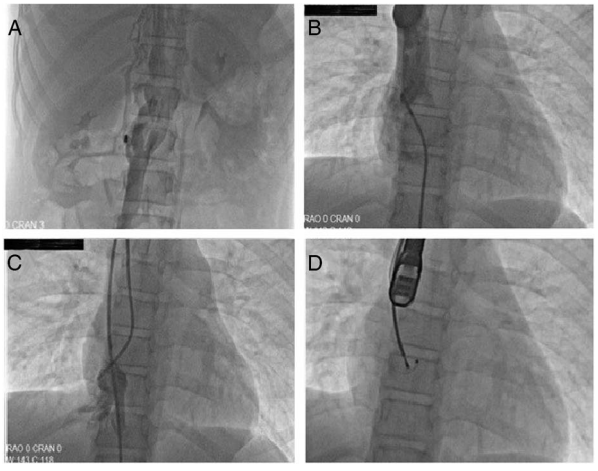

Figure 1.
The angiographic examination shows that interrupted inferior vena cava (a), continuation of inferior vena cava with azygos and superior vena cava, respectively (b,c), Amplatzer septal occluder delivered to the left atrium through the agilis steerable curved catheter (d).

P2747 - STENTING OF THE NATIVE RIGHT VENTRICULAR OUTFLOW TRACT IN THE SYMPTOMATIC INFANT WITH TETRALOGY OF FALLOT

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Objective: To assess feasibility, safety and effectiveness of right ventricular outflow tract (RVOT) stenting in symptomatic young infants with Tetralogy of Fallot (TOF).
Methods: Retrospective case note and procedure review of patients undergoing stenting of the RVOT over an 6 year period.
Results: Between 2010 and 2016, 10 patients underwent stent implantation; median age was 45 (range 1–364) days and median weight was 4 (2,2–7) kg. Median procedure time was 56,5 (30–233) min and fluoroscopy time 26,5 (12,6–77) min. There was one procedural emergency surgery. Two patients required ECMO support during the procedure. The pulmonary valve was deemed unsalvageable in all patients but one (median valve diameter 5 mm (range 4–8), median Z-score -3 (range -5 to -0,5). Saturations increased from 77% (50–85%) to 95% (85–98%) ($p < 0.005$). Three further catheter interventions were undertaken (balloon in 1, stent in 2). All patients underwent delayed surgery (complete repair in all of them) at a median of 140 (111–200) days post-stenting. No surgery was compromised by the presence of stents in the RVOT. There were no perioperative deaths. Median Nakata index increased from 80mm2/m2 (50–200) to 126 mm2/m2 (100–251)($p < 0.05$) before surgical repair.
Conclusions: In the symptomatic young infant with TOF, stenting of the RVOT provides a safe and effective management strategy, improving arterial oxygen saturation and encouraging pulmonary artery growth.

P2749 - TRANSCATHETER CLOSURE OF VENTRICULAR SEPTAL DEFECTS USING THE AMPLATZER DUCT OCCLUDER II DEVICE A SINGLE CENTER EXPERIENCE

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Background: Ventricular septal defects (VSD) may be closed surgically, by transcatheter approach or by hybrid procedures depending on the anatomy. We describe a single centre experience of transcatheter closure of VSDs using the Amplatzer Duct Occluder II (ADO II), aiming to evaluate the technical feasibilities and long term safety and efficacy.
Materials and Methods: Between Jan 2011 and Dec 2016, 281 patients between 8 months and 34 years, weighing 6-58 kg underwent percutaneous closure of VSD with ADO II at our institute. Patients with symptoms of left sided volume overload along with estimated Qp/Qs ≥1.5:1 were selected. The choice of ADO II device was based primarily on the size and location of the VSD, ease of the procedure either by arterial or venous approach of deployment, avoidance of an arteriovenous loop in majority of the cases, lower risks of post procedure atrioventricular block and the ease of retrieval in cases of accidental dislodgements. Post procedural echocardiogram substituted angiograms for evaluation of the position of the device, residual shunts, development of aortic regurgitation or tricuspid regurgitation.
Results: The location of the VSDs were: perimembranous -197 (70%), upper muscular- 71(25%), outlet - 11(4%), postsurgical residual- 2(0.7%). Right coronary cusp prolapse was present in 12 (4%) with no aortic regurgitation. In 2 patients, the device embolised but was successfully retrieved. The median duration of follow-up was 32 months. There were no incidences of heart block during the entire follow-up period. None of the patients had any aortic regurgitation post procedure. 2(0.7%) patients had developed mild tricuspid valve regurgitation.
Conclusion: Transcatheter VSD closure with ADO II device can be performed safely and successfully at all age groups with favourable results. In our study, there were no cases of heart blocks or aortic regurgitation. However, long term studies need to be ascertained for further follow up.

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P2755 - TRANSCATHETER PALLIATION IN PULMONARY VENOUS RETURN OBSTRUCTION

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Aim: The results of interventional cardiac catheterization for pulmonary venous return obstruction, present either before the surgical procedure or developed due to the procedure itself, were evaluated.
Material and Methods: Clinical and echocardiographic findings of all patients with congenital or acquired pulmonary venous obstruction, requiring transcatheter interventions, were retrospectively analyzed.
Results: Between 2014 and 2016 eight patients underwent nine catheter interventions to relieve the obstruction in pulmonary venous drainage. Five of the patients were male and 3 of them female. The median age was 3.2 months (between 7 days-13 months) and the median weight was 3.3 kg (between 2.6-6.5 kg). Among 8 patients, 5 had single ventricle and 3 had double ventricle physiology. All patients had total anomalous pulmonary venous connection (TAPVC) and five of them had right atrial isomerism with unbalanced complete atrioventricular septal defect. In 5/8 patients preoperative transcatheter vertical vein stenting and in 3/8 patients postoperative pulmonary vein stenting were performed to reveal the obstruction in pulmonary venous return. Only one patient required more than one intervention due to postoperatively developed pulmonary venous obstruction. Two of the patients had developed supraventricular tachycardia that was terminated by medication and one patient had cardiac arrest requiring cardiopulmonary resuscitation during the catheter intervention. There were no procedural mortality.
Discussion: The surgical mortality and morbidity of obstructed TAPVC either after or before surgery is high, especially in patients with low birth weight and unstable hemodynamic condition. Removal of the obstruction may improve outcomes for both preoperative and postoperative patients. Transcatheter stent implantation might be an alternative for treatment of obstructed TAPVC.

Table 1.

	Case 1	Case 2	Case 3	Case 4
Age	20 days	4 months	8 day	1 months
Weight (kg)	3.2	5	2.65	3.3
Gender	Male	Male	Male	Female
Diagnosis	RAI/CAVSD/DORV/PS/TAPVC/PDA/PVH	RAI/CAVSD/DORV/PS/TAPVC/PDA/PVH	TAPVC/PDA/PVH	RAI/CAVSD/DO/RVPS/TAPVC/PVH
Catheterization indication	Supracardiac TAPVC-obstructive	Supracardiac TAPVC-obstructive	Supracardiac TAPVC-obstructive	Supracardiac TAPVC-obstructive
Procedure	Stenting of vertical vein	Stenting of vertical vein	Stenting of vertical vein	Stenting of vertical vein
Vascular access	Right jugular vein	Right femoral vein	Left femoral vein	Left femoral vein
Procedural complications	No	Supraventricular tachycardia	No	Short term cardiac arrest
	Case 5	**Case 6**	**Case 7**	**Case 8**
Age	12 months	13 months	7 days	3.5 months
Weight (kg)	6.5	6	2.6	4
Gender	Female	Female	Male	Male
Diagnosis	RAI/CAVSD/PS/PVH (Glenn Shunt & (TAPVC repair)	RAI/CAVSD/DORV PVH (Glenn Shunt & (TAPVC repair)	TAPVC/ASD PVH	TAPVC-left PV-PVH (TAPVC repair)
Catheterization indication	Pulmonary venous obstruction	Pulmonary venous obstruction	Supracardiac TAPVC-obstructive	Pulmonary venous obstruction
Procedure	Stenting of PV & Azygo vein oclusion	1st Stenting of right & left PV 2nd Balloon angioplasty of right PV stent & stent implant tion into the left PV stent	Stenting of vertical vein	Stenting of left PV
Vascular access	Right jugular vein	Right femoral vein	Right femoral vein	Right femoral vein
Procedural complications	No	No	Supraventricular tachycardia	No

*RAI: Right atrial isomerism, DORV: double outlet right ventricle, CAVSD: Complete atrioventricular septal defect, PDA: Patent ductus arteriosus, PS: pulmonary stenosis, TAPVC: Total anomalous pulmonary venous connection, PV: pulmonary vein, PVH: Pulmonary venous hypertension

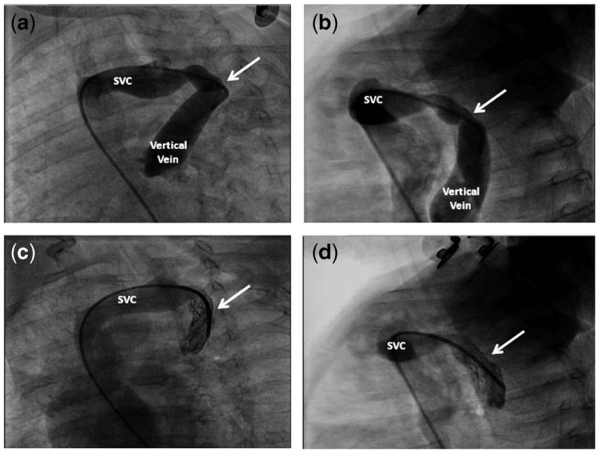

Figure 1.
a) Contrast injection into the collector sac, with emphasis on the narrowed part. b) Pull-back pressure recording from the collector sac to the vertical vein; c) Opening the stent, with emphasis on the stent waist; d) Contrast injection into the collector sac after stenting the vertical vein.

P2756 - APPLICATION OF AMPLATZER VASCULAR PLUGS IN VARIOUS CARDIOVASCULAR DISEASES AND CLINICAL CONDITIONS

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Backround: Amplatzer vascular plugs (AVP) are devices designed for the closure of extracardiac, high flow, medium to large vascular structures. The aim of this study is to evaluate the safety and efficacy of AVP's in the occlusion of vessels, defects and leaks at various localizations in several congenital cardiac diseases and clinical conditions.
Results: In the study, 3 to 21 years old 4 male and 7 female patients, to whom AVP's were applied, were included. Emergent application was required for the three of them, whereas the other eight were elective procedures. Our AVP indications were as

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follows: Aorto pulmonary collateral artery (MAPCA), pulmonary arteriovenous malformations (PAVM), azygos vein occlusion following the Norwood stage II operation, antegrade pulmonary flow occlusion following the modified Glenn operation, abnormal systemic artery and Scimitar vein occlusion in Scimitar syndrome, residual Swiss cheese ventricular septal defect (VSD) occlusion and aortic paravalvular leak (PVL) occlusion. The diameters of occluded vessels were ranging between 4.3 and 14.6 mm. Totally 18 AVP's were used for the 18 vessels: 13 AVP II (6 × 6-22 × 18 mm), 4 AVP IV (7 × 12.5-8 × 13.5 mm) and 1 AVP III (8 × 4 mm). Only one device was used for the closure of target vessel, defect or leak in each patient, with a 100% success rate. No complication was experienced during or after the applications.
Conclusion: There are a large variety of indications for AVP applications. They can be used safely and efficiently for the closure of vessels of different size and structure in various localizations in various cardiac diseases as an alternative to surgery. Complete occlusion can be achieved by a single device in large, abnormally shaped vessels, paravalvular leaks or residual ventricular septal defects.

P2779 - TRANSCATHETER CLOSURES OF SECUNDUM ATRIAL SEPTAL DEFECTS WITH ALTERNATIVE METHODS A SINGLE CENTER EXPERIENCE

Tugcem Keskin[1], Duran Karabel[1], Pelin Kosger[1], Ali Yildirim[2], Birsen Ucar[1], Zubeyir Kilic[1]
Eskisehir Osmangazi University, Department of Pediatric Cardiology, Eskisehir-Turkey[1]; Pediatric Cardiologist, Eskisehir-Turkey[2]

Introduction: The aim of this study is to emphasize successfull transcatheter closures of atypical ASDs with alternative methods.
Methods: 138 consecutive patients between 2010 and 2016 with significant ASD were considered for transcatheter closure with ASD occluder devices; 35 patients with inadequate septum or deficient rims were excluded from transcatheter closure after initial transoesophageal echocardiography.
Results: Procedure was successful in 89 cases (success rate 86,4%), among whom 69 had device deployment from the LUPV. 20 patients (7 male, 13 female; age 3 to 15 years, median 8 years; body weight 11–60 kg, median 25 kg) had deployments from the RUPV (8 patients) and LLPV (12 patients). One patient with interrupted inferior vena cava (IVC) had succesfull transjugular catheterisation closure, assisted with steerable introducer (Agilis NxT Steerable Introducer, St Jude Medical, Maple Grove, Minnesota). Six patients had successfull ASD closure with balloon-assisted technique and 2 patients had successfull ASD closure with Hausdorf sheat technique (Cook, Bloomington, IN) with deficient aortic rims. The diameter of defects and devices ranged between 10-28mm and 11-30mm, respectively of patients with atypical deployments. No severe procedure related complications occurred. At follow up (30 days to 5 years, median 3 years) complete closure was documented in all patients.
Conclusions: RUPV and LLPV techniques are usefull for transcatheter closure of ASD, also successfull closure of ASD can be managed by steerable introducer, Hausdorf sheat and balloon-assisted techniques in cases with deficient aortic rims or interrupted IVC.

P2783 - TRANSCATHETER CLOSURE OF LARGE AORTA TO RIGHT ATRIAL TUNNEL IN A NEWBORN

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Background: The aorta to right atrial tunnel is a rare cardiac defect described in infants, children and adults which has been treated most often by surgery. We describe the acute presentation and transcatheter treatment of a large defect in a newborn.
Case Presentation: A 2200 gram, 38 week, twin newborn was admitted to our facility with hemodynamic compromise related to a large aorta to right atrial tunnel measuring 6-8 mm in dimension. The defect was associated with torrential flow such that inotropic and prostaglandin infusions were required to augment systemic cardiac output. Echocardiography and CT angiography defined the anatomy as a posterior-type tunnel arising from the left coronary sinus of Valsalva communicating to the right atrium via an enlarged coronary sinus. The left coronary system was separate from the tunnel. A 3.5 mm restriction in the tunnel was noted posterior to the mitral valve and served as the target region for transcatheter device closure. The tunnel was entered from the aortic side and a 6 mm Amplatzer Vascular Plug 4 device was deployed in the region of vascular restriction posterior to the mitral valve. Flow persisted in the tunnel through the device mesh for several days, but inotropic and prostaglandin infusions were weaned off the day following treatment. Complete occlusion of tunnel flow was documented by echocardiography 6 days after device deployment. The patient developed right external iliac artery thrombus which resolved with 3 months of enoxaparin therapy. The infant was well and without cardiac symptoms six months following catheter treatment.
Conclusion: The aorta to right atrial tunnel is a rare congenital defect which may be amenable to transcatheter closure, even in small infants.

P2786 - TRANSCATHETER DEVICE CLOSURE OF DOUBLY COMMITTED VENTRICULAR SEPTAL DEFECTS

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Background: There remains international debate regarding efficacy and safety of transcatheter closure of doubly committed VSDs (DCVSD). This study reports a 5 year single centre experience with the approach.
Methods: Retrospective review, October 2009 to Jul 2014; 44 patients underwent device closure of DCVSD. Selection criteria: Weight >10 kg, no severe/moderate AR or cusp prolapse, defect <7mm, no other intra-cardiac abnormalities, plus evidence of pulmonary hypertension, left heart volume loading, or trivial/mild AR or cusp prolapse. Technique: Anterograde approach and AV loop, angiographic re-evaluation following sheath placement, echocardiographic evaluation of AR and outflow before and after device release.
Results: Median age 63 months (10-170), weight 18 kg (8-32), defect 3.7mm (2-6). Associated abnormalities: trivial to mild AR; 1 (2.3%), left heart dilation; 10 (22.7%), MR; 3 (6.8%), coronary cusp prolapse: 11 (25.0%). Devices used: PFM Coil: 10 (22.7%), ADO II: 21 (47.7%), PFM Coil and ADOII 13 (29.5%). Post-procedure murmur in 20: residual shunt disappeared on echo <48 hours in 18 (56,2%) the remainder after 6 months. Complications: haemolysis: 1 (2.3%) referred for surgery, embolization: 1 (2.3%), residual shunt 1 (2.3%) referred for surgery, RV outflow obstruction: 5 (11.4%) all resolved <3 months, AR increased: 4 (9.0%) 3 recovered to baseline <1 month, one referred for surgery. Mild LV outflow obstruction: 1 (2.3%) resolved <2 months.

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Conclusion: Device closure of DCVSD is a controversial but effective and safe option in selected patients. Coils are associated with a greater incidence of haemolysis than ADOII or Coil & ADOII. Close follow-up is required to identify haemolysis, outflow obstruction and outlet valve competence.

P2793 - TRANSCATHETER DEVICE CLOSURE FAILURE OF ATRIAL SEPTAL DEFECT ASSOCIATED WITH EUSTACHIAN VALVE

Tugcem Keskin[1], Pelin Kosger[1], Duran Karabel[1], Ali Yildirim[2], Birsen Ucar[1], Zubeyir Kilic[1]
Eskisehir Osmangazi University, Department of Pediatric Cardiology, Eskisehir-Turkey[1]; Pediatric Cardiologist, Eskisehir-Turkey[2]

Introduction: Atrial septal defect (ASD) is a common congenital heart defect which needs closure by transcatheter method or surgery. Device closure of ASD is a well-established procedure. We describe transcatheter closure failure of ASD associated with Eustachian valve.
Case Report: A 4-year-old girl was found symptomatic with functional class II. Physical examination revealed a healthy child with height 118 cm, weight 20 kg, pulse rate of 89 bpm, blood pressure of 110/72 mmHg, and respiratory rate was 22 per min. Cardiovascular system examination revealed normal first heart sound, wide fixed second heart sound. Chest X-ray showed mild cardiomegaly. Transthoracic echocardiogram demonstrated right atrial and ventricular enlargement with ASD secundum. Transesophageal echocardiogram measured a 17 mm ASD with 35 mm of total septum and adequate rims. Transcatheter closure was decided and performed with the patient under general anesthesia. Vascular access was obtained via the right femoral vein using 6 F sheath, right and left cardiac catheterization showed a pulmonary arterial pressure of 14 mmHg, Qp/Qs:1.8. The left upper pulmonary vein was entered using a 6 F multipurpose catheter. A 0.035" Superstiff wire was positioned and exchanged for a 9 F delivery sheath. A 22 mm Ceraflex septal occluder (Lifetech Scientific Corp., Shenzhen, China) was loaded on to the delivery system for the deployment from left upper pulmonary vein. Both discs were released however, right atrial disc could not be positioned well, TEE images then revealed a large Eustachian valve in right atrium. Several attempts were made to deploy the device, but were unsuccessful since Eustachian valve was slipped into right atrial disc and transcatheter closure of ASD failed.
Conclusion: Transcatheter closure of ASD in patients with large Eustachian valve may be challenging even with suitable size and adequate rims.

P2796 - TRANSCATHETER INTERVENTIONS IN CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES A CASE SERIES

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Background: Congenitally corrected transposition of great arteries (CCTGA) is a condition where the anatomic right ventricle (RV) assumes the function of systemic ventricle. The natural history of these patients are worse when associated with cardiac defects that increase the volume or pressure load on the already compromised systemic RV. We hereby present three patients of CCTGA with associated lesions who were successfully tackled by trans-catheter modalities.
Materials and Methods: GM, a 10 years old girl presented with NYHA II exertional dyspnoea. Echocardiography revealed CCTGA, an Ebstenoid tricuspid valve with moderate tricuspid regurgition(TR) along with a 7mm perimembranous ventricular septal defect (VSD). To unload the systemic ventricle it was decided to close the septal defect by percutaneous approach. Due to significant TR and massively dilated RV, the VSD was not delineated well on RV angiograms. The VSD was crossed from venous side using an NIH catheter and angiograms were taken in shallow RAO view with catheter tip just at the mouth of the defect. A 12 mm Lifetech Muscular VSD Device was then used to close the defect from the venous side. Post-procedure patient remained in sinus rhythm with no residual shunt and significant decrease in TR. BN, a 3 month old 5 kg baby with CCTGA had severe discrete post-subclavian coarctation of aorta with severe RV dysfunction and thus surgery was not preferred.The coarctation was successfully dilated using a 6mm x 2 cm Mini Tyshak balloon. AC, was a 8yrs old girl with CCTGA and a 3mm PDA. The PDA was closed successfully using a Flipper detachable coil.
Results: In all three patients, there were significant symptomatic improvement with increase in cardiac function and decrease in TR. All remained in sinus rhythm.
Conclusions: Any lesion increasing the volume or pressure load on the already compromised systemic RV in a CCTGA should be promptly corrected, to prevent further damage.

P2804 - EVALUATION OF BALLOON ANGIOPLASTY FOR AORTIC COARCTATION IN DIFFERENT AGE GROUPS A SINGLE CENTER EXPERIENCE

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Introduction: Pediatric patients with different age groups who underwent balloon angioplasty or stent implantation for aortic coarctation were evaluated.
Methods: From January 2006 to 2017, 69 patients with aortic coarctation underwent balloon angioplasty or stent implantation were evaluated. According to age at angioplasty, patients were divided into two groups: Group A (0-3 months, n = 21, 16 male/5 female, weight 2.5-6 kg) and Group B (>3 months, n = 48, 41 male/7 female, weight 6-68 kg).
Results: Mean peak systolic pressure gradient was reduced from 36,1 (15-58) mmHg to 8,4 (1-20) mmHg after balloon angioplasty [n = 21, 21 (100%) native] in group A and mean peak systolic pressure gradient was reduced from 40, 2 (17-74) mmHg to 10,6 (0-31) mmHg after balloon angioplasty or stent implantation [n = 48, 30 (62.5%) native, 14 (29.1%) recoarctation, 4 (8.4%) in-stent restenosis] in group B. There was no difference between groups for early success ($p < 0.001$). One patient required immediate surgery due to aortic dissection in group A. Mean follow-up period was 66 (1-120) months. There was no significant difference in terms of need for surgery between the groups ($p = 0.312$). Recoarctation developed in 9 (42.8%) patient after an average of 15 (0.5-60) months in group A, of which 7 required surgery, 2 required redilatation. Recoarctation developed in 18 (37.5%) patient after an average of 27(1-36) months in group B of which 9 required surgery, 9 required redilatation. Two (2.8%) patients had balloon angioplasty after surgical repair. 14 patients with coarctation underwent stent implantation, of those 4 had in-stent restenosis. During follow-up of 1-5 months, 3 (4,3%) patients died.

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Conclusion: Balloon angioplasty and stent implantation are usefull methods for aortic coarctation treatment. Patients should be monitored closely after procedures for recoarctation in all ages.

P2808 - EVALUATION OF SECUNDUM ATRIAL SEPTAL DEFECTS FAILURE RATES RESTRICTING FACTORS A SINGLE CENTER EXPERIENCE

Tugcem Keskin[1], Pelin Kosger[1], Duran Karabel[1], Ali Yildirim[2], Birsen Ucar[1], Zubeyir Kilic[1]
Eskisehir Osmangazi University, Department of Pediatric Cardiology, Eskisehir-Turkey[1]; Pediatric Cardiologist, Eskisehir-Turkey[2]

Introduction: The aim of this study is to emphasize the facts leading to transcatheter closure failures of ASDs or need for surgery.
Methods: 138 consecutive patients between 2010 and 2016 with significant ASD were considered for transcatheter closure with ASD occluder devices; 103 patients found suitable for transcatheter closure, 35 patients with inadequate septum or deficient rims were excluded from transcatheter closure after initial transoesophageal echocardiography.
Results: Procedure was successful in 89 of 103 cases (success rate 86,4%). Failure rate was 13,6% (14 cases, among whom 6 required surgery). Features of failed cases were; 5 inappropriate deployment despite several attempts due to malalignment ASD, 3 device embolization (surgery in 1, catheter retrieval in 2), 2 atrial septal anevrism, 1 Eustachian valve causing indentation into the right atrium resulting in improper positioning of device, 1 thrombus formation in right atrium before deployement, 1 failure despite Hausdorf sheath and balloon-assisted technique and 1 air embolism during prosedure. The diameter of defects and devices ranged between 16-24 mm and 17-30 mm, respectively of patients failed. Thirty five patients were found inappropriate for transcatheter closure (19 male, 16 female; age 2-17 years, median 7,6 years, body weight 14–64 kg, median 29 kg, defect diameter 16-32 mm) among whom 32 required surgery with deficient rims and inappropriate septums, 3 patients were thought to be evaluated for transcatheter closure in fallow-up.
Conclusions: Adequate total septum and sufficient rims, careful case selection, detailed imaging protocols and expertise in techniques are necessary for successful outcomes of transcatheter ASD closure.

P2814 - WHAT IS THE MAIN COUSE OF THE HEMI FONTAN FAILURE

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Introduction: Hemi-Fontan operation is one of the stages of treatment children with single ventricular types of congenital heart defects. Overall mortality after the operation is about 5%. The mail purpose of this study is to identify the most common causes of the failure after hemi-Fontan procedure.
Materials and Methods: Twenty nine children underwent cardiac catheterization during three month after hemi-Fontan procedure between 2014-2016. The results of these evaluations were analyzed for cause and effectiveness of the interventions.
Results: The majority of the study population were children with hypoplastic left heart syndrome (62,1%). Almost all children who underwent cardiac catheterization presented hypoxemia (89,7%) and symptoms of heart failure (72,4%), including fluids in the body cavities and generalized edema (24,1%). Other symptoms were decrease blood pressure in the lower part of the body (2 child) and superior vena cava syndrome (1 child). The most common cause of bad clinical condition after hemi-Fontan operation was left pulmonary artery (LPA) stenosis, which occurred in 79,3% of cases (23). Other causes include narrowing of the superior vena cava (3), stenosis of the distal aortic arch (3), stenosis of the right pulmonary artery (2), thrombus in hemi-Fontan system (3) and thrombus in left pulmonary artery (3). Left pulmonary artery stenting was performed in 18 children with good result in 17 cases (94,4%). Balloon angioplasty of LPA was effective in 53,8% of cases (7/13).
Conclusions: The most common cause of the hemi-Fontan failure is left pulmonary artery stenosis. Percutaneous stenting is an effective method of the pulmonary artery stenosis treatment.

P2834 - PERCUTANEOUS DUCT OCCLUSION IN VERY LOW WEIGHT CHILDREN A SAFE AND FEASIBLE ALTERNATIVE TO SURGICAL APPROACH

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Background: Patent Ductus Arteriosus (PDA) is the most frequent cause of heart failure in pre-terms and newborns with co-morbidities in neonatal intensive care units. Nowadays surgical closure is the treatment of choice in very low weight pre-terms but it carries significant morbidities and mortality risks. Catheter closure option can potentially overcomes these limitations. We present our experience in this population.
Materials and Method: From March 2012 to November 2014 we treated 12 newborns below 3Kg (mean weight 2.56, min 1.7, max 3.4 kg) with percutaneous approach. Amplatzer Duct Occluder (ADO) II and ADO II Additional Size were used.
Results: All 12 patients had hemodynamically significant PDA. Eight (66%) were preterms; comorbidities were severe broncodysplasia in 4 (25%), operated of aneurysm of Galeno vein in 1 (8.3%), congenital Rubella in 1 (8.3%), diaphragmatic hernia in 1 (8.3%) and other congenital cardiac defects (ASD and VSD) in 3 (25%). In eight patients (66%) medical therapy with indometacine was previously attempted but in four of them (50%) complications occurred. Eleven patients (92%) received furosemide, three (25%) ACE-Inhibitors therapy and two (17%) sildenafil. We register no mortalities nor morbidities due to catheterization. Nine (75%) received ADO II Additional Size and three (25%) ADO II. Mean fluoroscopy time was 15 minutes and mean contrast volume was 15 ml. No residual shunt at echocardiography was found in all cases. At follow-up (12 months) no deaths were registered. The heart failure therapy was stopped in all patients after procedure.
Conclusions: using currently available technology, percutaneous PDA closure in very low weight newborns is a feasible and safe alternative to surgical procedure. Early and mid-term follow-up is encouraging.

P2843 - PULMONARY ARTERY BANDING BALLOON DILATION PATIENT DESCRIPTION AND OUTCOME

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Pulmonary artery banding (PAB) is used for paliation of congenital heart defects with excessive pulmonary blood flow. The primary

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aim is to reduce pulmonary blood flow and protect the pulmonary vasculature from irreversible pulmonary hypertension. As the patient grows the obstacle becomes more restrictive causing reduced pulmonary blood flow and progressive cyanosis. In patients who are not candidates to corrective surgery, balloon dilation is an alternative to surgical intervention. We report our results with PAB balloon dilation (PAB-BD) over a ten-year period from 2007 to 2016. During this period 12 patients underwent PAB-BD. The most frequent diagnosis was aortic coarctation and ventricular septal defect(VSD) in 7 patients, followed by apical/multiple VSDs in 4 patients and a univentricular defect in one patient. The mean age at PAB was 58 days(3-238), and the mean post-operative gradient across the PAB was 65 mmHg(35-94), excluding the patient with univentricular defect (gradient of 33 mmHg). PAB-BD was performed at a mean age of 28 months (10-88) and was performed for severe cyanosis in five patients, restrictive/closed VSDs in five patients and severe RV hypertrophy in two patients. Balloons used were high-pressure in nine patients, low-pressure in two patients and in one patient a low-pressure balloon was used followed by a high-pressure balloon. The invasive gradient across the PAB decreased from a mean of 70 mmHg(35-124) to a mean of 46 mmHg(7-90). In patients with cyanosis there was an increase in saturation from a mean of 80,4% (60-87) to a mean of 89,9%(85-93). There were no complications of the procedure. None of the patients with small restrictive/closed VSDs required surgery. One patient died 10 years after the procedure. PAB-BD is a safe and effective procedure, improving saturation in cyanotic patients and avoiding the need for surgical pulmonary artery debanding and VSD closure in patients that evolve to small restrictive/closed VSDs.

P2846 - BALLOON ATRIAL SEPTOSTOMY IN THE CATHETERIZATION LABORATORY FACILITATES UMBILICAL APPROACH

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Introduction: Balloon atrial septostomy (BAS) is routinely performed under ultrasound control and can be performed via femoral (FV) or umbilical vein (UV). FVBAS requires a 7 F introducer for the Miller-Edwards septostomy catheter. FV thrombosis is common after FVBAS. Avoiding this complication is important as FV access facilitates future surgical or interventional procedures. Difficulty crossing the ductus venosus often makes UVBAS impossible. We investigate the use of fluoroscopy to facilitate UVBAS.
Methods: Patients undergoing BAS at <7 days of age by a single operator from 2010 to 2017 were included. Since late 2103 fluoroscopy has been used to facilitate UVBAS. Data were collected on location of procedure, access route, use of fluoroscopy, degree of urgency, radiation dose, and complications.
Results: During the study period a total of 52 BAS were performed. In the pre-fluoroscopy era (PreFE) 25 patients underwent BAS, 12 UVBAS (48%) and 13 (52%) FVBAS. In the post-fluoroscopy era (PostFE) 27 patients underwent BAS, 17 in the catheterization laboratory (63%). In 13/17 (76%) fluoroscopy was used to facilitate UVBAS. This was successful in 12/13 (92%). Fluoroscopy was available but unnecessary in 4 cases. In the one failure, fluoroscopy assisted diagnostic catheter passage via UV to right atrium but BAS was not possible. In the PoFE, UVBAS was successful in 21/27 (78%) and femoral in 6/21 (29%). Of the latter, UV was attempted unsuccessfully without fluoroscopy in 2. PoFE success rate of attempted UVBAS in the last 19 patients was 100%. Fluorography was used less often in urgent than in non-urgent UVBAS (3/9 vs 8/12 respectively). Complication rates were comparable between UVBAS and FVBAS. Fluoroscopy dose was negligible.
Conclusions: Since utilizing fluoroscopy, 78% of BAS have been performed using the UV compared to 48% prior to this, with no increase in complication rate and only minimal radiation.

P2853 - CATHETER BASE CLOSURE OF LARGE PDAS A CASE SERIES

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Back Ground: Patent Ductus Arteriosus (PDA) constitutes 6 to 11% of all heart defects. Surgical repair is a safe, widely accepted procedure with negligible mortality but it is also associated with morbidity, discomfort and a thoracotomy scar. Transcatheter closure of PDA is under continuous phase of evolution as far as technique and the closure devices are concerned. Multiple occluder devices have been studied and used for large PDAs of different shapes and sizes.We describe our single center experience with closure of large PDAs using the different devices depending on the anatomy, morphology and hemodynamics.
Materials and Methods: From July 2015 to December 2016, 25 patients (16 females,9 males) between 2 to 16 years underwent transcatheter closure of large PDAs (ranging from 6mm to 18 mm at PA end). On simultaneous measurements of PA and Aortic pressures, 4 children showed PA pressure >2/3rd of systemic pressure which further came down on oximetry, while the remaining 21 had PA pressure between ½ to 2/3rd of systemic pressure with features of reversibility.PDA delivery systems ranging from 6 F to 12 F were used.The devices used included Atrial Septal Occluder (2), Duct Occluder (14), muscular VSD device (5), vascular plug (1). 3 of the largest PDAs (12 mm ,16mm and 18mm) were closed with Post Infarct Muscular VSD Occluder measuring 20 mm,22 mm and 24 mm respectively.
Results: Transthoracic Echocardiography in immediate post intervention period revealed all devices in situ with no residual flow, aortic or branch pulmonary flow obstruction. Of 25 patients, 20 had moderate LV dysfunction which improved over next 2 to 3 days on inotropic supports, while 5 had mild LV dysfunction which improved eventually over 6 months.
Conclusion: Although surgical repair of large PDA is a safe, widely accepted procedure with negligible mortality, it is associated with morbidity, discomfort and a thoracotomy scar. Transcatheter closure of PDA is a reliable alternative to surgery.

P2858 - VALIDATION OF A US RISK SCORE FOR CARDIAC CATHETERISATION PROCEDURES IN A SINGLE CENTER PORTUGUESE PAEDIATRIC POPULATION

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Introduction: In 2016, the Congenital Cardiac Interventional Study Consortium (CCISC) developed and validated a scoring system that predicts the risk of serious adverse events

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(SAE's) for individual paediatric patients undergoing cardiac catheterisation (CRISP: Catheterization RISK Score for Pediatrics). We sought to validate this score in a Portuguese paediatric population and to compare its predictive accuracy with the original score.
Methods: A single-centre retrospective study, embracing patients under 18 years-old undergoing cardiac catheterisation, between January 2013 – January 2017. The area under the receiver operating characteristic curve (ROC) and Hosmer-Lemeshow test were used to assess the test performance and compare it with the original score.
Results: We included 459 patients in the study, median age was 6 years old, 47.3% female and 51,2% percutaneous interventions were performed. There were 17 SAE's (3.7% vs predicted 2.8%) with 0% mortality rate. All SAE's were amenable of immediate correction without morbidity. More experienced centers had SAE rate inferior than expected. The score exhibited good discrimination, with 0.79 area under the ROC curve (CI 0.66 to 0.91 vs original score 0.74) and an appropriate goodness of fit, with a Hosmer-Lemeshow test of 0.62.
Conclusion: The CRISP is useful risk stratification for SAE's in our patients undergoing cardiac catheterisation. Our outcomes are similar to other experienced institutions.

P2863 - TRANSCATHETER CLOSURE OF PERSISTENT FORWARD FLOW FROM THE VENTRICLE TO THE PULMONARY ARTERIES IN FONTAN PROCEDURES

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The Fontan procedure is a palliative surgery performed for the management of patients with complex congenital heart disease that are not amenable to complete biventricular repair. If excessive forward flow from the ventricle to the pulmonary arteries persists after the Fontan procedure, persistent pleural effusions, congestion of systemic veins or progressive ventricular dysfunction may occur. We report our series of transcatheter closure of forward flow from the ventricle to the pulmonary arteries in 5 patients after superior cavopulmonary shunt (n = 1) or complete Fontan procedure (n = 4). The main diagnoses were: double inlet left ventricle and transposition of great arteries (TGA); unbalanced atrioventricular septal defect, TGA and pulmonary stenosis (PS); tricuspid atresia and L-TGA; double outlet right ventricle (DORV) with unbalanced ventricles; and TGA and PS. Indications for closure were persistent congestion with edema in two patients, haemodynamic instability in the acute phase after Fontan procedure in one patient, persistent chylothorax in one patient (with cavopulmonary shunt) and low cardiac output with limited exercise tolerance in one patient. Mean age at the time of intervention was 10 years (1-20). In three patients an Amplatzer duct occluder was used and in two patients an Amplatzer atrial septal occluder. In two patients there was a small residual leak after the transcatheter procedure. No complications were reported. There was symptomatic improvement in all patients. The patient with haemodynamic instability and prolonged intubation was extubated the day after the procedure, two patients with edema reduced diuretics in the weeks following the procedure and one patient with chylothorax removed pleural drainage 5 days after the procedure. Transcatheter closure of ventricle-pulmonary artery communication is a safe and effective treatment for patients with residual flow after cavopulmonary shunt or Fontan procedure with early or late complications due to excessive pulmonary blood flow.

P2872 - ELIMINATING STENT SLIPPAGE USING A NEW COMBINED BALLOON SHEATH ASSEMBLY SYSTEM (NUDEL)

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Backgound: Percutaneous stent placement is used to treat vascular stenosis through previously placed long sheaths. Stents can slip off the balloon during its passage to the target. The stent must be retrieved, repositioned or remounted prolonging the procedure, some required surgical removal. Despite advances in technology and techniques, this complication persists. We describe our experience with a new method of stent delivery that eliminates stent slippage using an all-in-one stent-in-sheath balloon assembly system.
Material & Methods: The concept was to permit movement as one system (NuDEL, NuMed USA) minimising stent migration over the balloon. It evolved from the initial obturator shape system that was introduced through another short sheath, and did not allow free injection of contrast. The current NuDEL design overcame these drawbacks. The sheath has a hemostatic valve and permitted contrast injection. The BIB balloon has an obturator tip, using a 0.035" wire.
Results: From 2008, 21 patients (age 5.9-25.9 years) underwent 23 procedures to relieve pulmonary artery (PA) stenoses (n = 8), prior to pulmonary valve replacements (n = 12), PA conduit stenosis (n = 1) and coarctation (n = 2). The stent, front loaded on the balloon (8-22 mm) covered by the sheath (12-22 Fr). Pre-mounted stent system was used in 9 cases, covered CPTM stents in coarctation (n = 2) and for relieving proximal left PA stenosis while simultaneously occluding an aorto-pulmonary collateral. Multiple non-sustained arrhythmias (n = 8) occurred during the procedure. One obturator tip dislodged from the catheter. All stents remained on the balloon, and successfully deployed. In one patient where 2 stents were deployed to relieve lobar PA stenosis, one stent embolized proximally into the right hepatic vein, 12 hours after successful deployment.
Conclusion: Stent slippage is eliminated using the all-in-one balloon sheath assembly system, shortening of procedure time. It is ideal to rescue an unexpected arterial tear or perforation.

P2874 - HEART FAILURE IN A NEONATE DUE TO HEPATIC HEMANGIOMA DIAGNOSIS AND MANAGEMENT

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Hospital Evangelico, Pediatric Cardiologi, Cachoeiro De Itapemirim-Brazil[1]; Hospital Evangelico, Interventional Cardiologi, Cachoeiro De Itapemirim-Brazil[2]; Escola De Medicina Souza Marques, Medicine, Rio De Janeiro-Brazil[3]; Hospital Evangelico, Neonatologist, Cachoeiro De Itapemirim-Brazil[4]; Hospital Evangelico, Intervencionist Cardiologist, Cachoeiro De Itapemirim-Brazil[5]; Hospital Evangelico, Radiologist, Cachoeiro De Itapemirim-Brazil[6]; Hospital Evangelico, Cardiologist Surgery, Cachoeiro De Itapemirim-Brazil[7]; Sick Children, Pediatric Cardiologist, Toronto-Canada[8]

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Introduction: Hepatic hemangioma is a vascular tumor that involves arteriovenous shunt and which can culminate in heart failure in up to 50% of the cases. Death is the outcome in 70% of untreated cases. Cardiac symptoms are worse in younger patients.
Objective: To describe the clinical presentation, diagnosis, and management of a newborn with cardiomegaly, heart failure, anuria, and early hemodynamic instability.
Case Report: A full-term male baby weighing 3,950 g, with respiratory failure and cardiomegaly at chest X-ray, was submitted to mechanical ventilation, inotropic doses and peritoneal dialysis due to hemodynamic instability, tachycardia, signs of heart failure, hepatomegaly, edema, anuria and coagulation disorders. Echocardiography showed inferior dilation of the vena cava and heart chambers, moderate tricuspid insufficiency, and persistent ductus arteriosus. Abdominal ultrasonography revealed hepatic vascular alterations. Abdominal CT scan showed a huge hepatic hemangioma with arteriovenous shunt.The tumor occupied 80% of the liver, which prevented its resection. Due to the newborn's poor hemodynamic status, we decided to coil the tumor fistula by the percutaneous route when the baby was 12 days old. Soon after the procedure, angiography showed reduced shunt, and diuresis began. The coil was well positioned at the right hepatic artery as

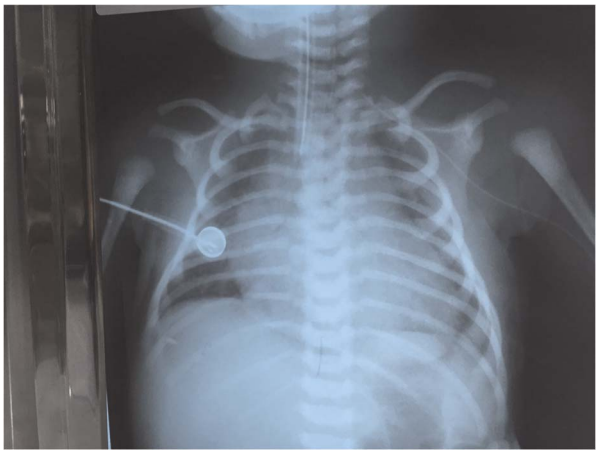
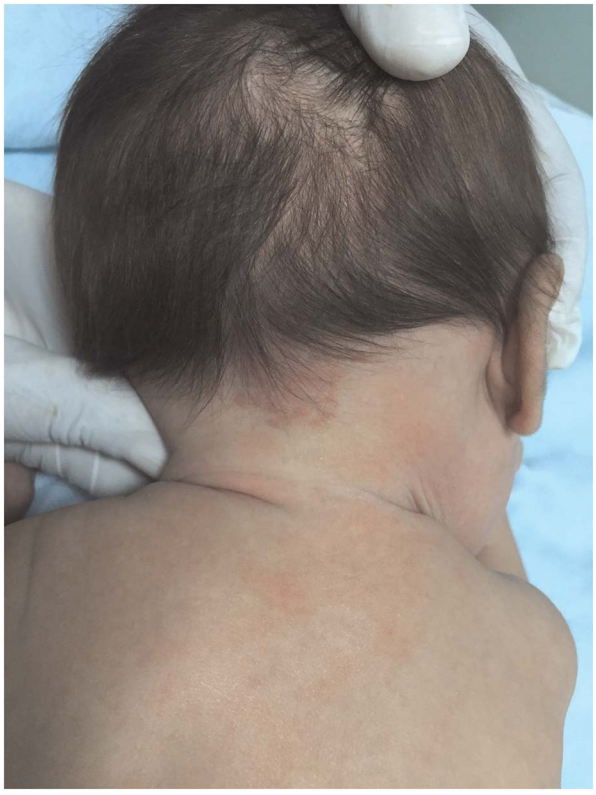


Figure 1.



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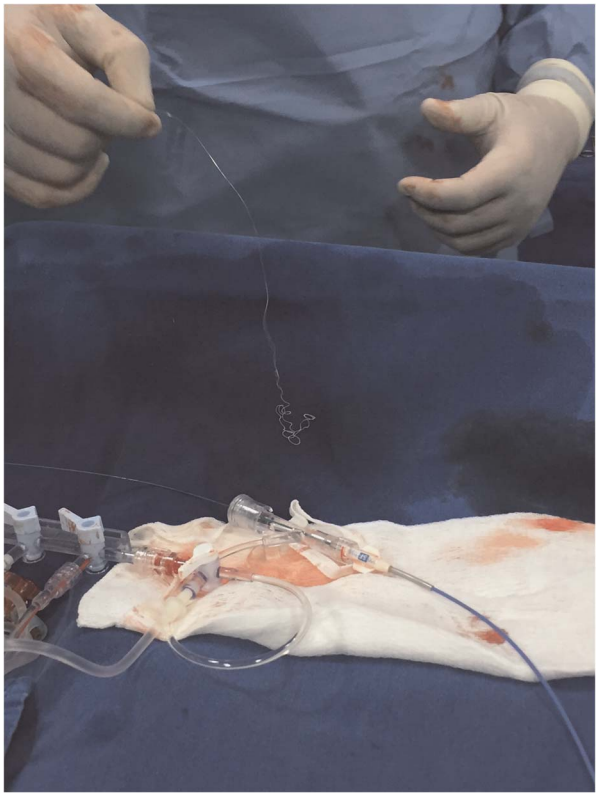

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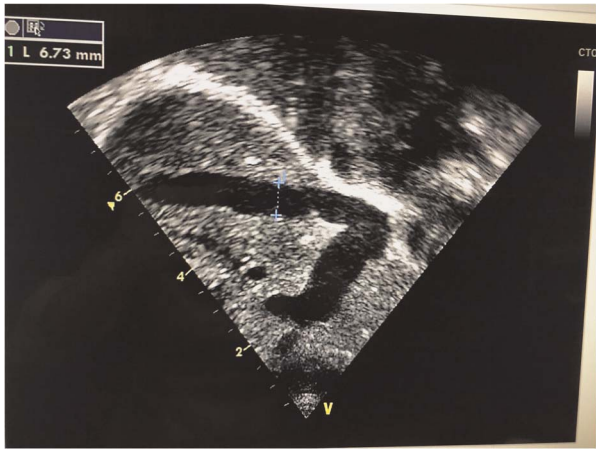

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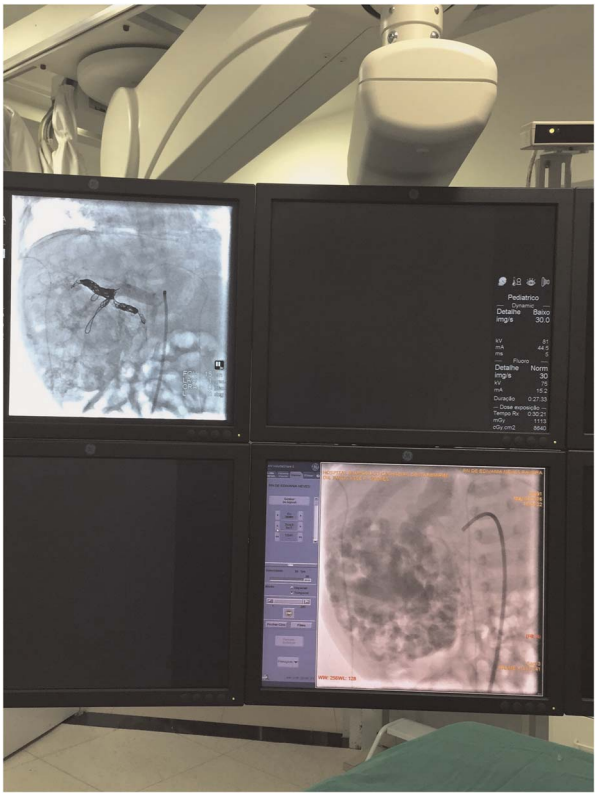

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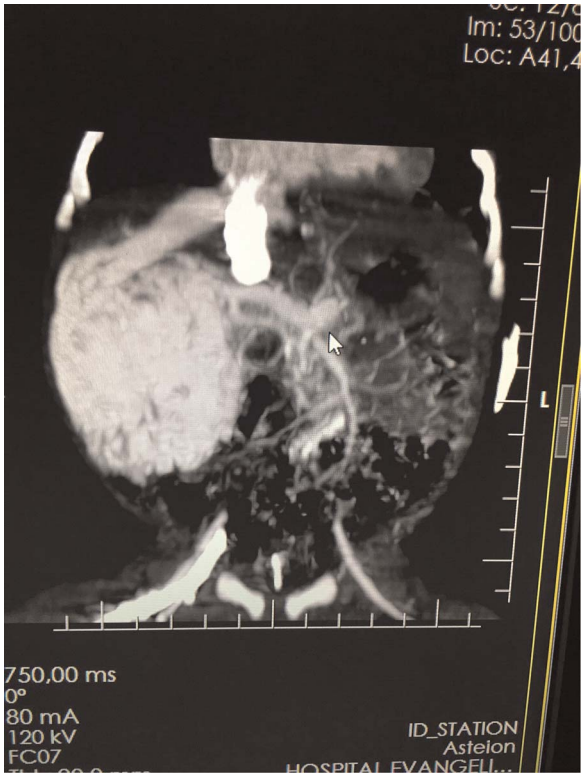
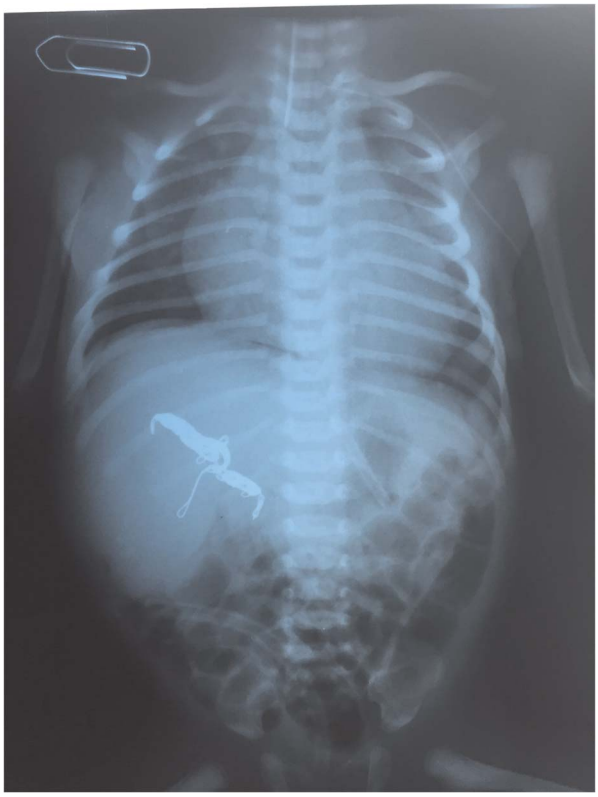


Figure 6.

revealed by post-procedure X-ray examination. Clinical conditions improved gradually, and the patient was discharged from hospital on propranolol. When the patient was six months old, he presented good cognitive and physical development and was being breast-fed. The latest abdominal ultrasonography, conducted at 4 months of age, showed liver with normal size and shape; there were scars in its central portion.

Conclusion: Although hepatic tumors are benign and tend to involute spontaneously, they may lead to heart failure and great morbi-mortality. Hepatic artery percutaneuos embolization was a challenging procedure that probably changed the progress and outcome in this case.



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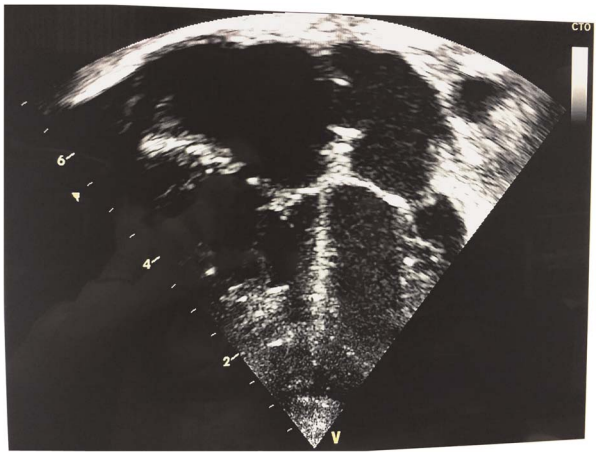

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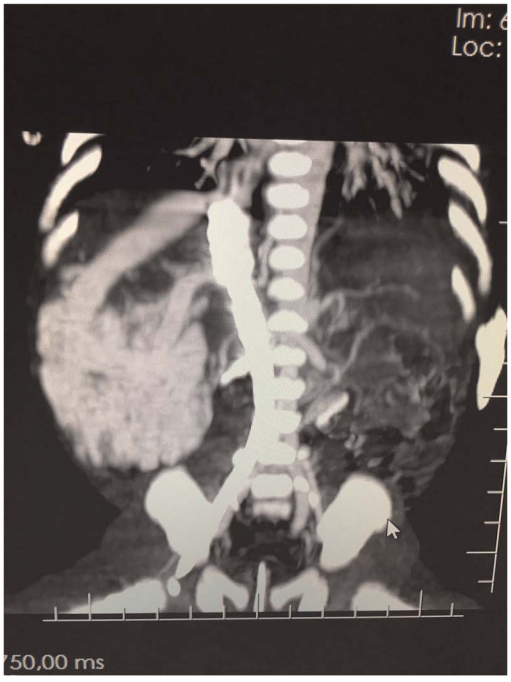

Figure 9.

P2880 - RESTENOSIS OF STENT IMPLANTED AORTIC COARCTATION DUE TO NEOINTIMAL HYPERPLASIA

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Introduction: Progressive neointimal proliferation may reduce the size of the stent lumen over time requiring redilatation.
Method: Balloon dilatation was performed in 3 male patients with stent implanted aortic coarctation due to instent restenosis.The age of stent implanted patients were 8, 11, and 16, respectively, post-procedural instent restenosis of patients detected in the second, first and third years of follow-up, respectively. Peak systolic pressure gradients measured before redilatation were 34, 35 and 52 mmHg respectively and the pressure gradients measured after redilatation were 2, 8 and 9 mmHg respectively. Mild proximal arch hypoplasia was present in 11 years old patient, restenosis detected in echocardiography at the first year of redilatation in the same patient requiring control angiography, pressure gradient was found lower than 20 mmHg, patient decided to be monitored with anhypertensive treatment, no redilatation prosedure was performed. Eight-year-old patient is in the 2nd year of redilatation and the 16 year-old patient is in the third year of redilatation, both are still followed with antihypertensive treatment.
Conclusion: Neointimal proliferation may cause restenosis in aortic coarctation. Redilatation is successful in treating restenosis caused by neointimal proliferation.

P2888 - PULMONARY ATRESIA AND INTACT VENTRICULAR SEPTUM – CAN WE STIMULATE THE RIGHT VENTRICLE GROWTH

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Background: Pulmonary atresia with intact ventricular septum (AP&IVS) is a defect which often requires single ventricle treatment. Prenatal pulmonary valve opening and valvuloplasty (FPV) is undertaken to preserve right ventricular size and function allowing for biventricular correction.
Materials and Methods: Echocardiographic examinations and postnatal follow-up of 64 fetuses with AP&IVS were reviewed. Patients were divided in two groups: diagnosed before 2010 year (n = 28). Basing on this results prospective study between 2011-2016 was conducted with the aim to prepare for FPV. There were 36 fetuses, including 9 who underwent FPV, 2 still in utero.
Results: The mean age of diagnosis was 28 + /-6 weeks in the first group, and 23 + /-5 weeks in the second. Heart defect was isolated in 85% of fetuses from 1stgroup and 72% from 2ndgroup.Chromosomal abnormality were in 4% and 14% in respective groups, other organs defects in 11% and 14%. Survival was 46% in the 1st group, only one patient achieved biventricular circulation. In the 2ndgroup survival rate was 66%. 7 patients after FPV have biventricular circulation, with one death after postsurgical complication. Right heart structures measurements during last prenatal exam (RV length, width, and TV annulus) expressed in Z-scores were compared between fetuses who underwent FPV and those who did not. They were significantly bigger in the FPV group: RV length: -1.26 vs. -5.74, RV width -0.41 vs. -4.62, TV annulus 0.81 vs. -2.43. In both groups pulmonary arteries were of normal size.
Conclusion: Prenatal diagnosis of AP&IVS improved with time. Retrospective analysis with majority of SV pathway enables to establish the national program for FPV. Right ventricle grows significantly after FPV. FPV enables biventricular pathway in majority of children.

P2893 - MID TERM FOLLOW UP OF MULTI CENTRE TRIAL OF THE OCCLUTECH® ACCELL® FLEX II SEPTAL OCCLUDER FOR TRANSCATHETER CLOSURE OF SECUNDUM ATRIAL SEPTAL DEFECTS

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Background: The Occlutech® Flex II ASD is currently used to close wide range of defects.Thrombus formation on atrial septaloccludersis rare, but it is a potential serious complication that could lead to an embolic event. A newer modification of this device (Occlutech® ACCELL® Flex II) has been designed to eliminate/reduce this potential complication. The aim of this study was to determine mid term follow up safety of this device.
Methods: A prospective, single-arm, open label, multi-center non-randomized pilot study in an application for CE mark. Thirty two patients were included in this study from a single center. Procedure protocol was approved by the institutional ethics committee. Patients were followed at 1, 30 and 90 days and six monthly post

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procedure. Color Doppler echocardiography was performed to quantify residual shunt (trivial shunt: jet width < 1mm;small shunt width 1-2mm; moderate shunt width 2-4mm and large shunt width > 4mm).
Results: Total number of patients enrolled were thirty two patients. 12 males(37.5%).Mean age was 7.5 ± 8.2 yearsand mean weight was 26 ± 23.9 Kg. All procedureswere done under general anesthesia and TEE was used to guide all procedures. Balloon sizing was performed in only three patients. All patients had adequate rims. The mean procedure time was 33.3 ± 14.7 minutes and mean fluoroscopy time was9.2 ± 7.8 minutes. The mean device size used was 18.5 ± 6.9 mm and the mean sheath size used was 10.8 ± 1.6 F. All patients had complete closure of their defects as demonstrated by TTE at 24-hour follow up. Mean follow up period was 3.2 years. No complications were encountered during the procedure or at follow up.
Conclusion: Mid term follow up of the Occlutech® ACCELL® Flex II showed it is a safe and effective device. The ACCELL coating on the device may enhance endotheliazation of the occluder and therefore may minimize the risk of thrombus formation.

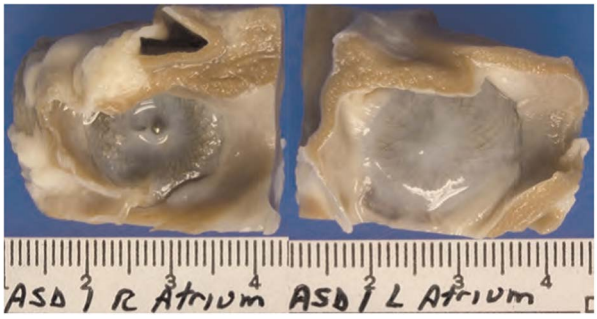

Figure 1.

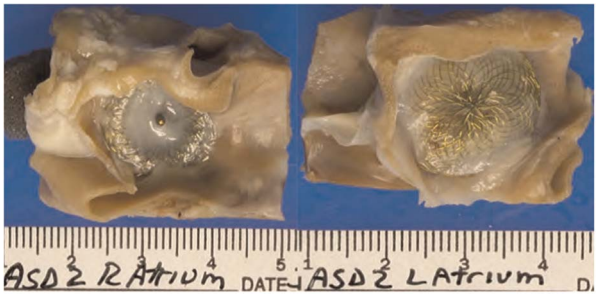

Figure 2.

P2899 - TRANSCATHETER CLOSURE OF SUPRACRISTAL VENTRICULAR SEPTAL DEFECTS – SHORT TERM OUTCOME

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Background: Surgery has been the mainstay for closure of supracristal ventricular septal defects (VSD). Transcatheter closure of supracristal VSDs have not been in vogue due to concerns of damage to the surrounding structures and outflow obstructions, risk of aortic incompetence (AI) from aortic valve damage. We present our series of 10 cases of device closure of supracristal VSDs, with a short term follow up of one year.
Materials and Methods: We followed the standard echocardiographic nomenclature for VSDs, whereby the VSDs which were well beyond the 12 o'clock position on the short axis views, were taken into this study. The VSDs were evaluated from other views as well, conforming to the diagnosis of supracristal VSDs. Cases where the RCC had summarily prolapsed into the VSD were rejected. 4 girls had whiff of preexisting AI, but their parents preferred device closure to open heart surgery, even after adequate counseling. Proper written consent was obtained in each of the cases in their vernacular language. The VSDs were best visualized in RAO 30 degree and Caudal 20 degree views, which we improvised for the supracristal VSDs. The VSDs were crossed from the aorta retrogradely and the ADO II devices were delivered from the aortic side. Meticulous echocardiographic evaluation of the devices were done after positioning them across the defect with special emphasis on the position, stability, possible impingement on the aortic valve and the pulmonary valve and any outflow obstruction.
Results: There was no residual flow, outflow obstructions or AI in any of the cases, on immediate and one year follow up examinations. The whiff of preexisting AI in 4 patients remained so even after a one year follow up.
Conclusion: Device closure of supracristal VSDs is possible in selected cases. Short term outcomes are encouraging, however long term outcomes need to be studied.

P2911 - DEVICE IN DEVICE FOR ASD CLOSURE

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Background: The atrial septum defect (ASD) is a common congenital heart disease. Its closure by percutaneous technique is safe, with a shorter hospitalization time, but it is not exempt of complications such as device malposition, thrombosis, endocarditis, residual shunt, arrhythmias, valvular regurgitation, cardiac perforation and sudden death.
Case report: 36-year-old woman with no previous comorbidities and an ASD, measuring 22 mm, occluded with a CARDIA ATRIASEPT 30 device, in August 2013. During follow-up she reported progressive fatigue and palpitations. Transthoracic echocardiogram (TTE) in March 2014 revealed residual shunt with a QP:QS of 2.3, which rised to 3 in November 2014 and 5.6 in a year later. These findings followed worsening of the symptoms. She underwent another catheterization in August 2016. On physical examination, she presented a splitting of S2 and mesosystolic murmur 3 + / 6 + in pulmonary and accessory aortic areas. During the procedure a fixed CARDIA device was detected in the ASD, with its metallic structure intact, but with complete loss of its polyvinyl alcohol membrane. Percutaneous closure of residual shunt was performed with multifenestrated AMPLATZER 35, successfully covering the previous prosthesis. This procedure was guided by three-dimensional transesophageal echocardiography. There was no residual shunt in the follow-up.
Discussion: Complications such as that described in this case are usually treated by surgical techniques. The occurrence of residual shunt through the central portion of the CARDIA ATRIASEPT

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prosthesis is still poorly described in the literature and its correction by the "intra prosthesis" technique has been shown to be a less invasive option with good results.

P2958 - TRANSCATHETER CLOSURE OF SUBARTERIAL VENTRICULAR SEPTAL DEFECT WITH THE AMPLATZER DUCT OCCLUDER II INITIAL CLINICAL EXPERIENCE

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Background: Surgical closure of subarterial ventricular septal defect (VSD) is well established but requires open heart surgery with cardiopulmonary bypass. This study aimed to evaluate the feasibility, safety and outcome of transcatheter closure of subarterial VSD with the Amplatzer duct occluder II (ADO II).
Materials and Methods: Between March 2014 and October 2016, a total of 12 patients with subarterial VSD who underwent transcatheter closure with ADO II were enrolled retrospectively. There were 7 males and 5 females. The age ranged from 2.3 to 58.6 years with the median of 7 years. The body weight ranged from 11.5 to 84 kg with the median of 26.3 kg. Echocardiography showed the VSD was supracristal subtype in 8 patients and intracristal subtype in 4. All patients had prolapse of right coronary cusp without subaortic rim. Mild aortic regurgitation was noted in 5 (42%) patients.
Results: Aortography showed VSD size ranged from 0.8 to 5.0 mm with the median of 1.9 mm. Device was successfully implanted in 92% (11/12) of patients. No complication was noted. Complete closure rate was 64%, 64% and 82% at 1-day, 1-month and 3-month follow-up respectively. Three (27%) patients developed new mild aortic regurgitation during follow-up.
Conclusions: The initial experience shows that transcatheter closure of subarterial VSD with ADO II is technically feasible, safe and effective. However, the influence of aortic regurgitation needs long term follow-up.

P2988 - USE OF SYMMETRICAL HYPERIOM PERI MEMBRANOUS VSDO FOR PERVENTRICULAR CLOSURE OF MUSCULAR SEPTAL DEFECT

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FCV, Cong Heart Dis, Bucaramanga-Colombia

The per ventricular closure of the muscular interventricular septal defects it's use in children with lightweight and it's incrementing with more promising results everytime. The devices used have been occlusors designed for muscular septum, which possess a length of 7 mm. We described a case which is treated with this closure mode through a new device designed for Perimembranous Ventricular Septal Defect in a 3 month old infant with 4,3 kg of weight and history of intrauterine growth retardation whom was diagnosed with the presence of muscular interventricular communication with length of 6 mm which is associated with the presence of aortic coarctation. It was taken to the surgery room and under general anesthesia a correction of aortic coarctation with terminal technical term extended by left lateral thoracotomy, sternotomy sequentially and interventricular communication approach per ventricular puncture was performed and a septal occlude device perimenbranous of 8 mm designed for interventricular communication was placed achieving a complete occlusion of the defect. The device used constitutes a feasible alternative in this patients and we consider the smallest waist length an advantage. Offering a more apt configuration further adapting the diameters of the interventricular septum in this age, which exposes less of the device's material towards the ventricular cavity.

P2993 - UNSATISFACTORY RESULTS OF CRITICAL AORTIC STENOSIS

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Introduction: Critical aortic stenosis is a rare disease in Koreans. The purpose of this study is to review the critical aortic stenosis cases experienced in a single center during the recent 12 years and to consider the issues to be considered when conducting treatment.
Methods: The period included in the study was from January 2005 to December 2016. We performed retrospective medical record review and image reanalysis for 7 patients.
Results: Seven patients were all male and the median age of first intervention was 11 days (2~15) and the average weight was 2.96 kg (2.4~3.3). The median follow-up period was 75 months (1-119). One patient died suddenly at home during the follow-up period. One patient underwent Ross operation and the remaining patients underwent balloon aortic valvuloplasty or surgical valvotomy. After first intervention, the aortic valve annulus increased by 0.47mm (0.2-0.8), but the coaptation height of aortic valve decreased by 0.44mm (-2.3-1.0). Prior to intervention, only two patients had grade I aortic regurgitation, but after intervention, aortic regurgitation was observed in all patients except one.
Conclusion: After critical aortic stenosis treatment, most patients have various degree of aortic regurgitation. These are mainly due to the prolapsed of the dysmorphic aortic valve leaflet and the decrease of the coapation height of aortic valve.

P2998 - UNDERSTANDING THE HEMODYNAMICS OF THE FETAL AND NEONATAL CIRCULATION AND ITS TRANSITION IN PULMONARY ATRESIA

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Background: Important changes in the circulatory system happen at birth, when gas-exchange function is transferred to the lungs. Modifications of the system ensue for adaptation and survival of the newborn. Patients with pulmonary atresia (PAt) who have a ductus-dependent pulmonary-circulation are at risk as they may fail to make adequate transition to postnatal life. In PAt there is a left-to-right flow through the ductus arteriosus (DA), contrary to normal fetal circulation and most cardiopaties. Since few models have evaluated the hemodynamic effects of the transition to postnatal life, the objective of this study was to develop mathematical models of the circulation of healthy and PAt infants during fetal and postnatal life to understand the changes between these four conditions.
Methods: Lumped parameter models (LPM) of the cardiovascular circulations of healthy-fetal, PAt-fetal, healthy-newborn and PAt-newborn were developed. The main assumption was that oxygen supply and blood flow to vital regions was maintained between

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models. Models were validated with hemodynamic variables from healthy infants found in literature.
Results: We developed realistic LPM of fetal and neonatal circulations of both healthy and PAt infants, which allow estimation of flow properties that cannot be assessed from clinical measurements and to study the effects of the transition from fetal to postnatal life. The models give point-to-point pressure and flow estimates in all four conditions, confirm the fetal right-heart and newborn left-heart dominance in the healthy models and the importance of DA flow in PAt infants (Table 1).
Conclusions: The proposed LPM are good approximations to assess hemodynamic changes in fetal and neonatal circulations and their transition for healthy and PAt patients. Further development of the model can be useful for assessing the importance of isolated parameters and for simulating the effect of surgical and device interventions to repair congenital heart defects in PAt patients.

Table 1. Stability conditions for each model.

		Fetal (F)			Neonatal (N)		
					CO healthy: 230 mL-min^{-1} kg^{-1}		
		CCO: 540 mL-min^{-1}kg^{-1}			CCO PAt: 460 mL-min^{-1} kg^{-1}		
		Pressure drop, ΔP [mmHg]	Flow, Q [mL-min^{-1} kg^{-1}	Distribution of CCO, %	Pressure drop, ΔP [mmHg]	Flow, Q [mL-min^{-1} kg^{-1}]	Distribution of CO/ CCO, %
Heart hy (H)	Upper systemic circulation	47,9	151,2⋆	28,0%	67,6	151,2⋆	65,7%
	Foramen oval	0,7	113,4	21,0%	Closed		0,0%
	Pulmonary circulation	51,5	102,6^{+}	19,0%	20,7	102,6⋆⋆	100%
	Ductus arteriosus	2,8	221,4	41,0%	Closed		0,0%
	Lower systemic circulation	47,9	270,0	50,0%	67,6	270^{++}	27,2%
Pulmonary atresia (PAt)	Upper systemic circulation	41,9	151,2⋆	28,0%	41,5	151,2⋆	32,9%
	Foramen oval	7,5	437,4	81,0%	8,3	437,4	50,0%
	Pulmonary circulation	49,3	102,6^{+}	19,0%	49,8	102,6⋆⋆	50,0%
	Ductus arteriosus	0,0	102,6	19,0%	0,0	102,6	50,0%
	Lower systemic circulation	41,9	270,0	50,0%	41,5	270^{++}	13,6%

Model Assumptions:
⋆ Blood flow to the upper systemic circulation is equal in all models.
+ Blood flow to the pulmonary circulation is equal in HF and PAtF.
⋆⋆ Blood flow to the pulmonary circulation is equal in HN and PAtN.
++ Blood flow to the lower systemic circulation is equal in HN and PAtN.

Notes:
-CCO: Combined Cardiac Output
-CO: Cardiac Output

P3000 - HYBRID PULMONARY VEIN STENTING IN THE PATIENTS WITH REFRACTORY TO SURGICAL PULMONARY VEIN STENOSIS REPAIR

Jakyoung Yoon[1], *Gibeom Kim*[1], *Eunjung Bae*[1], *Kyungjin Ahn*[1], *Mikyung Song*[1], *Chungil Noh*[1], *Woonghan Kim*[2], *Jaegun Kwak*[2], *Jeong Ryul Lee*[2]
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Back ground: Pulmonary vein stenosis (PVS) is still frustrating disease with an extremely high mortality, especially in the patient with multiple severe PVS. Hybrid pulmonary vein stenting is a rescue treatment for recurrent and malignant PVS. We performed intraoperative hybrid pulmonary vein stent placement successfully in the 4 patients who had been operated multiple times for severe PVS after total anomalous pulmonary vein return (TAPVR) repair.
Cases and Review: Four patients were identified between 2013 and 2016, who were diagnosed with PVS and underwent hybrid pulmonary vein stenting. We performed hybrid PV stent placement on one patient who had progressive multiple PV stenosis and severe pulmonary hypertension despite recurrent surgical PV widening after cardiac type TAPVR repair. After hybrid stenting with coronary drug eluting stent (DES) and surgical pulmonary vein angioplasty, pulmonary vein stenosis was relieved and pulmonary hypertension was improved. Other two patients was diagnosed mixed type TAPVR (supra-cardiac + cardiac) and had severe multiple PVS despite recurrent surgical repair. These patients underwent twice hybrid pulmonary vein stenting with bare-metal stent (BMS) and pulmonary vein ballooning. Last patient who was diagnosed as functional single ventricle with pulmonary atresia and supra-cardiac type TAPVR developed progressive severe PVS after surgical repair of TAPVR. This patient also underwent hybrid stent placement. Hybrid pulmonary vein stenting resulted in sufficient relief of pulmonary venous stenosis to permit clinical stabilization in all patients. All patients received aspirin and clopidogrel after the operation. Three patients had undergone several elective further catheterizations for pulmonary vein ballooning or large stent insertion after hybrid stenting procedure.
Conclusion: We consider hybrid pulmonary vein stenting is to be a useful treatment modality for recurrent PVS patient to achieve longer freedom of restenosis than repetitive surgical PV widening.

Table.

	Case 1 (F/46 months)	Case 2 (M/20 months)	Case 3 (M/ 19 months)	Case 4 (M/ 13 months)
Original disease	Cardiac type TAPVR	Mixed type TAP VR	Mixed type TAPVR	Supra-cardiac tvpe TAPVR, DORV with pulmonary atresia, unbalanced AVSD,
Number of operation Before hybrid stent	3	4	3 1 Including hybrid procedure (PV balloon)	3
Age at hybrid stenting	14 months	12 months	9 months	12 months
PVS degree	RUPV stenosis LPV os obstruction RLPV severe stenosis	RLPV 2mm os stenosis RUPV/ LUPV/LLPV 1mm	RUPV/RLPV 1mm LUPV 2mm	RUPV/RLPV os stenosis Diffuse small LPV
Pulmonary vein stent and diameter⋆	Atrial fenestration LPV(45mm DES)/ RLPV (4mm DES) RUPV: cut-back angioplasty	Atrial fenestration LUPV (6mmBMS)/ LLPV(6mm BMS)/ RUPV balloon(6mm)/ RLPV(7mm BMS)	Atrial fenestration RUPV(6mm BMS)/ RLPV balloon (7mm)/ LUPV(7mm BMS)	RUPV: cutback angioplasty RLPV(7mm BMS/, LPV stent(7mm BMS)
Follow-up duration after hybrid procedure	33 months	9 months	11 months	2 months
Number of further procedure	4 interventions	1 intervention 1 hybrid LUPV LLPV: balloon (6mm) RUPV (6mm BMS)	4 interventions	-

⋆DES: drug-eluting stent, BMS: bare metal stents

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P3006 - ROUTINE TRANSCATHETER CLOSURE OF PATENT DUCTUS ARTERIOSUS IN PREMATURE BABIES

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Background: Few previous studies have reported the feasibility of transcatheter closure of patent ductus arteriosus (PDA) in premature babies. The use of the AMPLATZER DUCT OCCLUDER II ADDITIONAL SIZES (ADO II AS) seems potentially interesting in this patient population. However, the type and size of PDA that can be percutaneously closed with this device in premature babies remains unknown.
Materials and Methods: Since December 2015, 26 premature babies (10 male) underwent transcatheter closure of their PDA in 3 institutions. The last 18 cases were performed consecutively between October 2016 and January 2017 since this procedure has been offered as a first line treatment, regardless of the patient's weight as well as size/type of PDA. With the exception of the 2 initial cases who underwent an additional 3-French arterial access, all catheterisations were accomplished through a 4 French femoral venous access only. The ADO II AS was used in all cases, mainly under echocardiographic guidance.
Results: The median age and weight of patients at the time of transcatheter PDA closure was 29 days (range 8 to 100) and 1250grams (range 823 to 2900 grams). The median diameter of the PDA was 3.5 (1.5 to 5)mm. The ADO II AS was used in all cases. There was one failure, no immediate complications and no residual shunts. During follow-up, no aortic obstruction was demonstrated. Two months after the procedure one patient experienced severe left pulmonary stenosis requiring surgery. Two other patients had mild left pulmonary stenosis with an increased Doppler velocity of 1.6 and 2 m/s, respectively.
Conclusions: Based on this preliminary experience, routine transcatheter occlusion of PDA in premature babies with the ADO II AS is safe and effective. Longer follow-up is needed and larger studies must be performed to confirm that this procedure can be an interesting alternative to surgery.

P3012 - RIGHT HEART CATHETERIZATION USING ANTECUBITAL VENOUS ACCESS IN PATIENTS WITH COMPLEX CONGENITAL HEART DEFECTS

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Introduction: The antecubital venous approach is a technique that provides very high comfort of both patient and physician, which is now forgotten but easy to access. In this report, we present the technique and features of the patients with antecubital fossa vein approach.
Technique: The most prominent antecubital fossa vein punctured with a 21-gauge needle. A 0.021 guide-wire was inserted through the needle into the vein. After seeing the tip of the wire in the vena cava superior with free movement, a 4-5 Fr sheath was inserted into the antecubital vein. All manipulations performed by using right Judkins catheter and 0.035-inch hydrophilic guide-wire. Heparin was not routinely given and the sheaths were pulled out at the end of the procedure using manual compression.
Patients: Between January 2014 and December 2016, we performed right cardiac catheterizations through the antecubital fossa veins of nine patients with various clinical indications. Six patients (67%) were male and three patients were female. The median age was 14, 6 years (3, 5 to 24 years) and the median weight was 43 kg (12 to 80 kg). Right arm was used in five patients and left arm in four. All patients except one had complex congenital heart disease with Glenn anastomosis. One patient was operated for sinus venosus ASD, abnormal pulmonary venous return and had total occlusion of VCS. In the five of the patients femoral approach was used concomitantly for additional diagnostic reasons. In two patients abnormal veno-venous collaterals occluded with various devices. Patients whom the diagnostic angiography performed discharged after 3-6 hours.
Conclusion: The antecubital venous approach can be performed easily and safely for diagnostic and theuropatic catheterisation in patients with complex congenital heart defects.

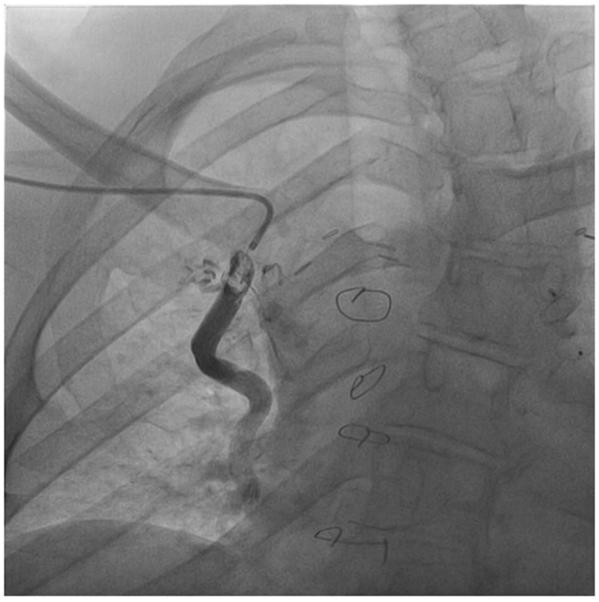

Figure 1.

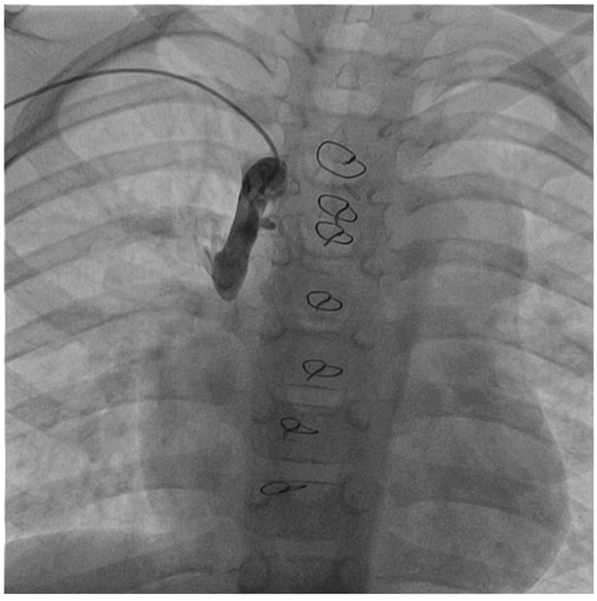

Figure 2.

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P3040 - LATE STENT REVASCULARIZATION OF ISOLATED PULMONARY ARTERY OF DUCTAL ORIGIN

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University of Tennessee Health Science Center, Pediatric Cardiology, Memphis-United States[1]; University of Colorado-Denver, Cardiology, Aurora-United States[2]; University of Colorado-denver, Pediatric Cardiac Surgery, Aurora-United States[3]; University of Colorado-denver, Pediatric Cardiology, Aurora-United States[4]

Background: We report our experience with stent revascularization (SR) of isolated pulmonary artery of ductal origin (IPADO) prior to surgical repair.
Methods: SR involved use of a 4Fr catheter, microcatheter and 0.014" hydrophilic coated wire to traverse occluded ductal tissue with subsequent coronary stent implantation.
Results: Between 5/08 and 5/16, 7 subjects had attempted SR-IPADO. Median age = 12mo (1-61.5), median wt = 8.4 kg (2.7-17.6) and 4 had right IPADO. A premature neonate was kept on PGE for 1 month until SR where the ductus was patent but severely stenotic (1.1mm). Median pulmonary arterial resistance was 5.0units (2.8-8.6). SR was successful in 4/7. A remnant aortic ampulla was present in all and a ductal "beak" (Fig A) leading to the occluded ductus was present in all successful (S) SR but a blunt ending ampulla (Fig D) was found in 2/3 unsuccessful (U) SR. The IPADO distal mean diameters for S-SR increased from average 3.4mm ± 0.64 to 8.9mm ± 0.85 ($p < 0.001$) (Fig B,C) prior to surgical repair. There was significant diffuse branch PA stenosis in 2/7 (1 S-SR; 1 post mBT shunt) and 2/4 S-SR developed in-stent stenosis all requiring intervention prior to surgery. Therapy for mild-moderate PHTN was used in 4/4 S-SR prior to corrective surgery. Surgical repair occurring at median 12.5mo (range 5-19) post SR involved interposition graft in 3 (9, 10 and 14 mm diameter) and 1 direct anastomosis. Surgery was facilitated by having a "virgin" chest. In median follow-up of 39.5mo (6-88) post surgery, only the subject with diffuse PA stenosis remains on therapy for PHTN. 2/3 U-SR remain unrepaired. Pulmonary vein wedge 3DRA imaging/roadmapping facilitated SR in 4/7 (Fig E). Complications of the SR-IPADO procedure include femoral artery thrombus (1/7) and occlusion (1/7).
Conclusion: Stent re-vascularization of IPADO is safe and when successful provides adequate growth of the effected artery and facilitates surgical correction.

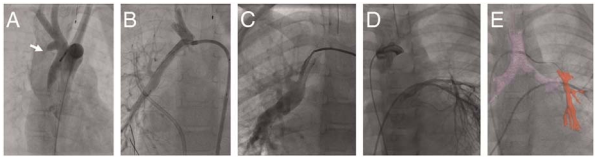

Figure.

NURSING

P1186 - DEVELOPMENT AND IMPLEMENTATION OF A PEDIATRIC CARDIAC INTENSIVE CARE NURSE RESIDENCY PROGRAM NURSE COORDINATORS AND CLINICAL EDUCATORS

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Background/ Hypothesis: A sustainable, reproducible Nurse Residency Program (NRP) may address the gap between nurses' academic preparation and their clinical practice in the Pediatric Cardiac Intensive Care Unit (PCICU). However, in order for a NRP to be reproducible and sustainable local expert nursing staff require mentoring to become NRP coordinators and clinical educators.
Materials and Methods: International nursing experts, in collaboration with Children's HeartLink, developed a three one-week NRP through an interactive peer review process including specific coordinator and clinical educator training. Three six-hour coordinator/educator classes focused on teaching/learning methods in both the classroom and in the clinical setting. Training included, but was not limited to, facilitating small group learning, critical thinking, instructional technology, promoting active learning, and concept mapping. Topics that focused on the clinical aspects included the preceptor role, mutual goal setting, providing meaningful and supportive feedback, evaluation conferences, and gaining acceptance of the clinical educator role in the ICU. Coordinators were invited to biweekly teleconferences to discuss program and facility planning issues, as well as to provide ongoing support, coaching, and encouragement.
Results: Six hospital nurses attended the 3 clinical educator sessions with two being prepared for the NRP coordinator role and four as clinical educators during initial NRP implementation. The NRP was subsequently implemented by coordinators and clinical educators with less support from Children's HeartLink volunteers. Some nurses from the leadership group were no longer involved during the second NRP session due to staff turnover or promotion highlighting the need to train adequate local staff in order to sustain the NRP.
Conclusions: A NRP can be reproduced in a resource constrained environment by local expert nursing staff if there is sufficient training and support for selected nursing staff at the onset of the NRP.

P1358 - REGIONAL BRAIN GRAY MATTER VOLUME LOSS IN ADOLESCENTS WITH SINGLE VENTRICLE HEART DISEASE

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Background/Hypothesis: Adolescents with palliated single ventricle heart disease (SVHD) show autonomic, mood, and cognitive deficits, indicating brain changes induced functional issues in the condition. Scattered brain injury appears preoperatively and postoperatively in both gray and white matter areas in SVHD. The purpose of this study was to examine regional brain gray matter volume changes in SVHD over controls using voxel based morphometry procedure.
Materials and Methods: Regional whole-brain gray matter volume changes were examined using voxel based morphometry procedure. Two high-resolution T1-weighted images were collected from 18 SVHD adolescents (age: 15.7 ± 1.1 years, male: 10) and 31 healthy controls (age: 16.0 ± 1.1 years, male: 17) using a 3.0-Tesla magnetic resonance imaging (MRI) scanner. Whole-brain high-resolution T1-weighted images were realigned and averaged, gray matter tissue type partitioned, normalized to a common space and modulated, smoothed and compared between groups using ANCOVA (covariates: age and gender; corrected threshold; $p < 0.05$; voxel size, 696 voxels).

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Results: No significant differences in age ($p = 0.35$) or gender ($p = 0.96$) emerged between groups. Multiple gray matter sites, including the bilateral thalamus, caudate, putamen, insular cortices, and left hippocampus showed volume loss in SVHD compared to controls [all regions $p < 0.05$](Figure 1).
Conclusions: Adolescents with palliated SVHD show significant gray matter volume loss in areas that regulate autonomic, mood, and cognitive functions. The pathological mechanisms contributing to gray matter volume loss may include developmental and hypoxia/ischemia induced processes operating in the condition.
Grant Support: This research work was supported by National Institutes of Health R01NR013930 and R01NR016463. Figure 1 Caption: Gray matter loss in brain areas including (a) caudate nucleus, (b) thalamus, (c) putamen, (d) left insula lobe and (e) left hippocampus, in SVHD patients compared to controls.

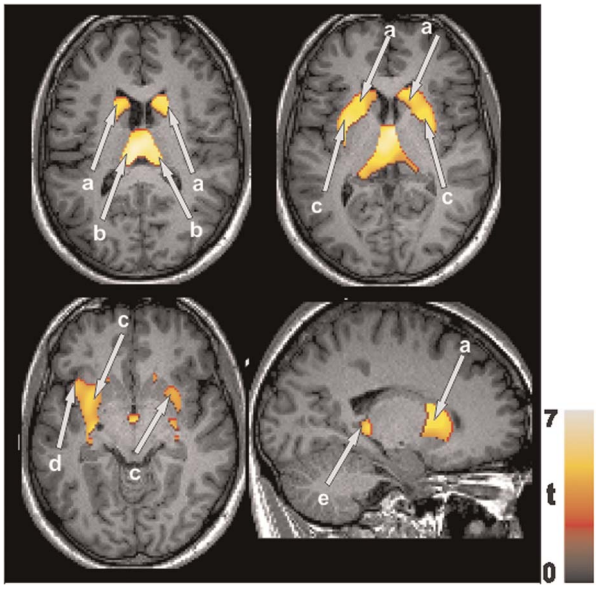

Figure 1.

P1434 - HEMOSTASIS WITH NEW DESIGNED BAND AFTER PEDIATRIC CARDIAC CATHETERIZATION

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Backgrounds: Physical restraint after cardiac catheterization make a patient confused. We have used adhesive plaster bandages for hemostasis on which peeling off made patients feel pain. Additionally glue-related dermatitis was significant events. Since 2014 hand-made girdle band, to press groin for hemostasis was introduced and used in preparation. Purpose: To evaluate effect of "Maki-chan", for appropriate pressing the puncture site and reducing dermatitis and pain. Objects: Patients over three years old who received pediatric cardiac catheterization in our ward.
Methods: We manually made a new designed girdle band of cotton with a piece of Velcro and rubber. The band cross just above the puncture site and rubber attached to cotton band can keep effectively press the groin. Before the procedure patients tried it on and recognized fitting and painless. The puncture site was disinfected and dressed by gauze with small adhesive plaster. The folded gauze with about 2 cm thickness compressed the puncture site and Maki-chan was worn on. From medical record during last four years hemostasis effects were compared between large adhesive plaster band and "Maki-chan", with small adhesive plaster.
Results: Large adhesive plaster bandage was used in 119 cases during 2 years. Maki-chan with small plaster was in 113 case during 2 years. No bleeding occurred in both group. Some patients in large adhesive plaster group showed localized dermatitis. Discussions: New band with non-stretchable cotton and folded gauze compress the puncture site effectively. Reducing plastered area could decrease pain and dermatitis. Trying band on before procedure is expected to be effective for preparation, even though just looking at. Preparation with trying band on helped reduction of confusion after procedure.
Conclusions: New band can reduce pain and dermatitis. Preparation with it can lead child's cooperation and strengthen hemostasis.

P1485 - SOCIOCULTURAL BELIEFS AND SOCIAL PRESSURE IN EVERYDAY LIFE OF CHILDREN LIVING WITH CONGENITAL HEART DISEASE IN THE PALESTINIAN WEST BANK

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Background: In Palestinian territories, approximately 700 newborns are diagnosed with congenital heart disease (CHD) every year. The advances in early diagnosis, treatment and postoperative care have resulted in increased survival rates. Research focus has shifted from survival to long-term follow up, wellbeing, daily life experiences and psychosocial consequences. Much remains to be learned about everyday experiences of children with CHD in occupied territories since challenging social conditions can have an impact on management and support. The aim of this study was to explore the everyday experiences of children with CHD and of their parents living in the Palestinian West Bank.
Material and Method: We adopted a qualitative descriptive design and analyzed the data using content analysis. A purposeful sample of children aged 8-18 years with CHD ($n = 9$) and their parents ($n = 9$) was selected. All participants were interviewed individually.
Results: Facing barriers and managing challenges was the overall theme that emerged and it consisted of four themes; socio-cultural burden and destiny, challenging physical/structural limitations, self-perceptions and concerns about not standing out and restraints in health care due to the political situation. Facing barriers represented the struggle children and their parents experienced which affected their daily life. Daily life was influenced negatively by socio-economic restraints due to political struggle, societal stereotyping and children's perceptions of their illness. Some considered the illness as a fate and therefore found comfort in their religious beliefs.
Conclusion: The negative impact of sociocultural beliefs and social pressure in everyday life of children with CHD and their parents is related to insufficient knowledge and misunderstanding about CHD in the Palestinian society. In addition, the political situation also contributes to a challenging daily life for children with CHD.

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P1555 - SELF REPORTED HEALTH STATUS IN ADOLESCENTS AND ADULTS WITH CONGENITAL HEART DISEASE COMPARED TO HEALTHY CONTROLS

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Background/Hypothesis: Improved life-expectancy in adolescents and adults with congenital heart disease (CHD) comes with need for continued follow-up. Self-reported health status is vital information for ongoing follow-up and clinical decision-making. The Individual's perceived health living with CHD may be related to disease severity. The purpose of this study is to examine self-reported health status in adolescents and adults with repaired or palliated CHD, compared to healthy controls.
Materials and Methods: Cross-sectional, comparative study; CHD participants were recruited from two cardiac centers in Los Angeles and controls from high schools and the community. Self-reported health status was measured using the Short-Form Health Survey (SF-36v2) which includes 8 subscales and two summary scores [physical and mental health scores 0-100; mean 50 ± 10; worse health status <50]. Demographic and clinical data was extracted from medical records. Descriptive and non-parametric statistics, including Kruskal–Wallis test were used to assess CHD severity [single ventricle (SV) and biventricular (BV)] and control group differences.
Results: 303 participants [CHD = 150 (SV = 87, BV = 63)/Controls = 153], ages 14-53 years. Within the CHD group, 55% male, 58% had a single ventricle, 77% had undergone $\geq$ 2 cardiac surgical procedure. CHD group reported significantly lower perceived health status than healthy controls in both physical and mental health [median 47 vs. 58, $p < .001$; 48 vs. 57, $p < .001$], respectively (see Table). SV group reported worse perceived physical and mental health compared to the BV group. Both CHD severity groups had lower mental health. All SF-36 subscales were statistically significant between groups [$p < .05$] except for bodily pain.
Conclusions: Adolescents and adults with SV and BV CHD had significantly lower self-reported health status compared to healthy controls. Interventions are warranted that focus on improving both physical and mental health status in this high-risk population.

Table 1. Perceived Health Status Score [SF-36 scores] in Single and Biventricular CHD Compared to Controls

SF-36 Scales	CHD Total N = 150 Median (IQR)	Healthy Total N = 153 Median (IQR)	BV CHD N = 63 Median (IQR)	SVCHD N = 87 Median (IQR)	P Value†
Subscales					
Physical Function	75 (60-95)	100 (95-100)	76 (65-95)	71 (60-90)	<.001*
Role Function Physical	75 (56-100)	100 (94-100)	72 (50-100)	73 (56-100)	<.001*
Bodily Pain	84 (62-100)	84 (74-100)	84 (64-100)	80 (51-100)	.271
General Health	62 (42-77)	80 (67-90)	68 (54-80)	52 (35-72)	<.001*
Vitality	58 (43-73)	63 (50-75)	62 (48-75)	54 (38-69)	.004*
Social Function	81 (62-100)	100 (75-100)	87 (50-100)	74 (63-100)	<.001*
Role Emotional	83 (50-100)	92 (75-100)	75 (50-100)	83 (58-100)	<.001*
Emotional Health	70 (60-85)	80 (70-85)	70 (60-75)	75 (55-85)	.030*
Summary Scales					
Physical Health Total	47 (42-55)	58 (55-60)	50 (45-56)	46 (41-51)	<.001*
Mental Health Total	48 (40-56)	57 (47-60)	46 (40-55)	45 (40-51)	<.001*

SF-36 = Short Form Health Survey; CHD = Congenital Heart Disease; BV = Biventricular; SV = Single Ventricle †P values represent non-parametric Kruskal-Wallis Test comparison among 3 groups [BV, SV and Controls]. *Statistically significant ($p < 0.05$).

P1641 - REFINED ANALYSIS THE CRITICAL EARLY WARNING SCORE FOR CHILDREN WITH CONGENITAL HEART DISEASE AFTER CARDIAC SURGERY

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Objective: To divide the level the critical early warning score and time interval to predict the prognosis of children with congenital heart disease after cardiac surgery.
Methods: 607 cases who was living in CICU after cardiac surgery was chosen to make a score by the electronic scoring system once an hour until the children died or transfer out CICU. According to the different outcomes, the children were divided into diffient groups. ROC curve, AUC, Youden index, Hosmer-lemeshow test was used to choose the optimal critical value and diagnosis the effect of the scoring.
Results: ① I-no risk:The mean score was 0 to 2 points and the highest score was 0 to 5 points, the highest score occurred after transferring to CICU about 3-4 days, 2 days after the highest score roll-out CICU. ② II-mild critically ill:The highest score was 6 to 10 points,and the mean score was 3 to 4 points keeping the trend and dynamic score, corresponding to the end for the critically ill. ③ III-moderate critically ill: The mean score was 5 points, the highest score was 11-18 points and keeping the trend of dynamic score is children, dead end, in death risk of interval ④ IV-severe critically ill: The mean score was 6 to 7 points, the highest score was 19 to 39 points, the highest score occurred after transferring to CICU after 2.5 days, 4 hours after result in rescue.⑤ V- extremely critical: The mean score was 8-39 points, the marking, the highest score was 40-57 points and peak appears in the CICU after 2-3 days, 4 hours after the peak,and ending for rescue invalid death.
Conclusion: The 5 risk levels of critical early warning score will help CICU nursing staff to be sensitive early-warning prompt on time and to take the countermeasures.

P1642 - VERIFY THE ELECTRONIC ILLNESS SCORING INFORMATION SYSTEM AND THE PAPER VERSION SCORING TOOLS FOR CHILDREN WITH CONGENITAL HEART DISEASE AFTER CARDIAC SURGERY

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Objective: To verify the degree of consistency and efficiency between the electronic illness scoring information system and the paper version scoring tools for children with congenital heart disease after cardiac surgery.
Methods: Two raters using two kinds of evaluation tools 50 cases of children with congenital heart disease surgery in CICU for measuring time each score used at the same time. Intraclass correlation coefficient (ICC) was used to analyze the two scoring tools' consistency degree; Independent samples t-test was used to analyze timeliness.
Results: ① The two scoring tool high degree of consistency: the same raters using two scoring tool for children with the same score the consistent rate of raters respectively A: 94%, B: 96%. ICC were 0.993 (0.988 0.996), 0.990 (0.983 0.995), $P < 0.01$. Two raters used the same kind of tools for children with the same score consistent rate respectively was: print score tools, electronic scoring system, 90% and 100%. ICC were 0.984 (0.972 0.991), 1 (1, 1). ② The timeliness validation results showed: paper score tool

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54.48 ± 5.93, 5.93 ± 5.28 (s); electronic scoring system: 21.26 ± 3.11, 3.11 ± 3.86 (s), P < 0.01. Both A and B raters used paper score tool ,the time has the difference: respectively 54.48 ± 5.93, 5.93 ± 5.28 (s), P < 0.05; Both A and B raters used electronic scoring system to make a score,and there was no statistically significant difference,the time was: 21.26 ± 3.11, 20.92 ± 3.86 (s), P > 0.05.
Conclusion: The electronic scoring system highly restored the paper version scoring tool, with higher accuracy and less affected by human factors, and it can avoid the artificial errors, In terms of timeliness advantages: a score of time saved nearly 2/3.

P1661 - IMPLEMENTING SAFE PERFUSION PRACTICES IN THE KARDIAS CONGENITAL HEART SURGERY PROJECT IN MEXICO

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Introduction: Since 2007 the World Health Organization proposed the implementation of safety checklists for all periods of the surgical processes. Considering congenital heart surgery, a special area to be considered is extracorporeal support, also denominated perfusion. We describe the implementation of a pre-perfusion checklist, immediate post perfusion report and a perfusion handoff protocol to the cardiac intensive care unit in the KARDIAS project in Mexico.
Material and Methods: Moment 1: A three phase checklist (as advised by the IQIC project) was integrated to the perfusion record to asses safety prior going into cardiopulmonary bypass.
Moment 2: A report sheet including the main details occurring during the perfusion period was designed. This format is analyzed both by the surgeon and the perfusionist before they leave the operating room. This document includes very concise information including: bypass and aortic clamping duration, water balance, low flow duration, etc. and it's included in the patient's record.
Moment 3: We also designed a postoperative perfusion hand-off protocol, which included a report sheet containing details occurred during perfusion, such as: mean NIRS values, mean hematocrit value during the procedure and hypothermic circulatory arrest duration, but what we consider of utmost importance is the inclusion of an adverse incident report section. This synthetic report is read by the perfusionist in charge of the surgical procedure, in between the surgeon's and anesthesiologist's segments at the hand-off process. Finally this document is included in the medical record of the patient.
Results: We present the three perfusion formats implemented in the KARDIAS project. We also show the perceptions concerning effective communication and process fulfillment of the surgical nurse participating in the handoff process, the receiving intensive care unit nursing staff, anesthesiologists, surgeons and intensive care physicians.
Conclusions: Considering the overall team opinion we propose the implementation of a safe perfusion practice strategy in order to enhance quality.

P1706 - CONTINUING EDUCATION PROGRAM IN THE HOSPITAL SETTING QUALITY IMPROVEMENT INITIATIVE

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Background: Nurses involvement in quality improvement is limited by specific challenges, related to traditional nursing education that does not always adequately prepare them for their evolving role in today's contemporary hospital setting. Healthcare demands require from educators to use novel teaching frameworks to enhance learners' competency in the clinical settings.
Purpose: To assess the efficiency of the case-based education program in improving the level of knowledge among acute nurse practitioners in the intensive care unit (ICU).
Methods: 33 ICU nurses (the mean age 39 ± 6 years) were included in the study from June, 2015 to July, 2016. The mean total years of the ICU services - 11 ± 6 years. The pretest-posttest model was used to assess the efficiency of the hybrid educational program. Both groups underwent post-test II after each educational phase to assess the retention of knowledge. All nurses were enrolled into 2 groups according to the results of the pre-test. Subjects who gave >50% of the correct answers were included in Group 1 (n = 11) and underwent a lecture-based education. Subjects who received <50% were included in Group 2 (n = 22) and underwent case-based tutorials based on virtual patient.
Results: Both groups showed significant improvement in scores between the pre- and posttests (Group 1 -56.64 ± 3.17; Group 2 - 56.50 ± 0.70) with similar knoweledge retention after 1 year (Group 1 - 54.45 ± 2.42; Group 2 - 55.0 ± .000). No statistically significant difference in mean test scores could be found between 2 groups at each point: 1.4% (95% CI –0.6% to 3.4%) for post-test I, and –0.3% (95% CI –3.9% to 3.3%) for post-test II.
Conclusion: Case-based education using virtual patient provides the potential to teach nurse practitioners with greater flexibility using different learning methods addressed to different learning domains than classroom instruction.

P1763 - PREVENT REGRESSION IN FUNCTION AFTER EARLY CARDIAC SURGERY HELP FIND A WAY

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Background/Hypothesis: The early years (0-5), are recognized as the period of greatest brain vulnerability to adverse events, including those stemming from early complex cardiac surgery. While limited, the research also indicates that children following complex cardiac therapies are at risk of ongoing adverse neurocognitive, behavioral, emotional, and academic outcomes as they progress into their school years. We set out to 1) determine the extent to which regression occurs in pre-academic and academic functioning after life-saving therapies at <6 weeks of age and to 2) develop

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a practical, workable framework of intervention to prevent regression in the child's developmental trajectory.
Method: We determined a mean lack of expected progression of functional skills and a regression of skills (loss of ability) using the parent-completed Adaptive Behavioral Assessment System II (domain, mean + sd, 10 + 3; delay = < 4). Focus Groups consisting of health care professionals, allied health workers and parents engaged in active dialogue about prevention of regression under the sub-topics of: at the bedside, in follow-up, early childhood, school-readiness, and school age.
Result: Pre-academic and Academic scores were 8.9(3.1), 8.2(3.4) and 6.4(3.3) at 2, 4,and 8 years of age respectively. The proportion of children with delay increased, 4.1%, 9.7%, 32% at these ages relative to peers. The Focus Groups yielded well organized and helpful approaches to intervention.
Conclusion: Lack of progression and regression of skills occurs in children after complex cardiac therapies. Evaluation and intervention for functional deficits for these children requires collaborative care early on in the child's life. This study informs our developing framework to enable all children to have support both before and after school entry. Functional abilities are a very important consideration in developmental evaluations of children assessed for school readiness.

P1766 - IMPACT OF PARTNERSHIP WITH INTERNATIONAL DATABASE AND CARDIOLOGY AND PEDIATRIC CARDIOVASCULAR SURGERY CENTER IN BRAZIL

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Introduction: In Brazil, there is a 65% deficit of congenital heart defects surgical correction. Improving quality of care in Brazilian pediatric cardiovascular surgery services would be facilitated by the establishment of parameters and objective data to evaluate surgical results offered.
Objective: To analyze the evolution of quality of care for children with congenital heart disease after the establishment of the partnership between the International Quality Improvement Collaborative Database (IQIC) for Congenital Heart Surgery in Developing Countries and a Center for Cardiology and Pediatric Cardiovascular Surgery in Brazil.
Methods: Data collection from a single center of cardiology and pediatric cardiovascular surgery in Brazil between 2011 and 2015 independently and with external audit in partnership with an international database. The data include preoperative information, as well as postoperative information.
Results: In the preoperative period, there was a difference in patients with altered nutritional appearance between the years (P = 0.004). There was also a difference regarding mechanical ventilation and inotropic therapy (P = 0.006). In the postoperative period, surgical procedures classified in RACHS-1 risk category show a small variation between groups (P = 0.445), with categories 2 and 3 prevailing. The number of surgical site infections reduced from year to year (P = 0.130), but it was evident bacterial sepsis and other major infections had a significant reduction (P = 0.023 and P = 0.026, respectively). The 30-day postoperative follow-up evolved satisfactorily with a discrete reduction in mortality for both in-hospital (P = 0.172) and 30 day-mortality (P = 0.156).
Conclusion: The five-year analysis showed a positive evolution in the quality of care for children with congenital heart disease after the establishment of the partnership with the IQIC database, mainly in the areas of infection reduction and mortality.

P1849 - VALIDATION OF AN EARLY WARNING SCORING TOOL FOR IDENTIFYING PEDIATRIC CARDIAC AND NON CARDIAC PATIENTS AT RISK FOR CARDIOPULMONARY ARREST

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Background: Most inpatient pediatric arrests are preventable by early recognition and treatment of clinical deterioration. Children with cardiac disease have the highest arrest rates however early warning scoring systems had not been validated in this population. The Cardiac Children's Hospital Early Warning Score (CHEWS) was designed and implemented to identify clinical deterioration in this high risk population. The CHEWS was then applied to all inpatients outside of the intensive care units (ICU). Nurses assess patients' CHEWS scores during routine vital signs and follow an escalation of care algorithm: routine care (score 0-2), increased assessment/intervention (3-4), or ICU consult/transfer (≥5).
Objective: This study s objective was to validate the CHEWS in identifying clinical deterioration in cardiac and non-cardiac patients using the previously validated Brighton Pediatric Early Warning Score (PEWS) for comparison.
Methods: A retrospective cohort study reviewed all non-ICU patients at a quaternary academic pediatric hospital. Specificity, sensitivity, area under the receiver-operating characteristic curves (AUROC) and early warning times were calculated for the CHEWS and PEWS in patients that experienced an arrest or unplanned ICU transfer (n = 424) and a randomly selected comparison cohort (n = 1026).
Results: see attached table
Conclusions: CHEWS has excellent discrimination to identify clinical deterioration in cardiac and non-cardiac pediatric patients. CHEWS had a higher AUROC and sensitivity compared to PEWS. CHEWS provided a longer early warning time the PEWS for clinicians to treat patients experiencing clinical deterioration.

Table.

	Cardiac Cohort (n = 312)		Non-Cardiac Cohort (n = 1136)	
	CHEWS	**PEWS**	**CHEWS**	**PEWS**
AUROC*	0.917	0.785	0.902	0.798
Sensitivity score (score ≥3)	95.3%	54.7%	91.4%	73.6%
Specificity (score ≥3)	76.2%	86.3%	67.8%	73.5%
Median early warning time for score ≥3* (hours)	9.25	2.25	11.1	3.8
Median early warning time for score ≥ 5 * (hours)	2	0	8.5	0.6

Validation of CHEWS and PEWS for identifying deterioration in cardiac and non-cardiac patients (*p < 0.001).

P1888 - EFFECT OF AN EDUCATIONAL INTERVENTION ON NURSE'S KNOWLEDGE ABOUT LOW CARDIAC OUTPUT SYNDROME IDENTIFICATION AND MANAGEMENT

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Objective: To determine the effectiveness of a two hour teaching module in increasing pediatric cardiac intensive care nurses knowledge about low cardiac output syndrome (LCOS) identification and management.
Methods: A 2 hour teaching module consisting of a didactic lecture, small group discussion (on case scenarios) on identification and management of pediatric LCOS was designed by an MScN student in collaboration with pediatric critical care physicians. Twenty six cardiac intensive care nurses were included in the study. Their knowledge on LCOS was tested before and after this education intervention. Data was collected on a structured proforma and knowledge was assessed on a self-developed validated tool. Paired T test was used to determine the difference in knowledge.
Results: Mean age of the participants was 35 ± 15 years, 20 participants were males, two were nursing intern, 18 were registered nurses and 6 were critical care technicians, 12 of particiapnts had no formal critical care qualification. Four participants had <1 year experience, 11 had 1-5 years' experience, 5 had 6-10 years' and 11-15 years' of critical care experience. Twenty six participants were basic life support certified and 5 were pediatric advanced life support certified. Mean score of participants increased from 58.41 ± 12.5 to 81.2 ± 18.4 (p value <0.05).
Conclusions: On job focused small group educational intervention is very helpful to increase the knowledge of critical care nurses which can then translate into good patient outcomes.

P1933 - LOW MOLECULAR WEIGHT HEPARIN ADMINISTERED BY SUBCUTANEOUS CATHETER IS A SAFE AND EFFECTIVE ANTICOAGULATION REGIMEN IN SELECTED INPATIENT INFANTS AND CHILDREN WITH COMPLEX CONGENITAL HEART DISEASE

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Background/Hypothesis: Intravenous therapeutic unfractionated heparin (IV UFH) is the first-line anticoagulant agent in infants and children with complex congenital heart disease admitted to the hospital. Disadvantages of IV UFH are unpredictable pharmacokinetics requiring frequent phlebotomies and the need for continuous intravenous access. Low molecular weight heparins administered by a subcutaneous indwelling catheter device (SC LMWH) might be an effective alternative.
Objective: To compare the efficacy and safety of SC LMWH with IV UFH in this patient population in a case control study.
Materials and Methods: Demographic data, number of needle punctures, anticoagulation parameters and complications of patients admitted to the pediatric cardiology inpatient unit and requiring invasive anticoagulation were recorded.
Results: Data are depicted in the Table. There was no difference between age, gender, underlying anatomic diagnosis, and the number of treatment days per patient between groups. The number of needle punctures was significantly lower in the SC LMWH as compared to the IV UFH group. Most activated partial thromboplastin times obtained in the IV UFH group were not within therapeutic range, whereas almost half of the SC LMWH group had therapeutic anti-Xa levels. Neither group had a bleeding or thrombotic complication. Minor and transient local reactions, such as redness or a small hematoma, occurred in 46% of patients of the SC LMWH group; there were no major complications, such as bleeding at the site or infection requiring antibiotics.
Conclusion: As compared to the current gold-standard of IV UFH, SC LMWH offers a safe anticoagulation regimen for inpatient infants and children with complex congenital heart disease providing more efficient therapeutic anticoagulation and a reduction in needle punctures, thus causing less pain and anxiety in this pediatric population.

Table.

	IV UFH	SC LMWH	p-value
Nr. of patients (males)	42 (21)	13(5)	
Nr. of treatment days	102	49	
Age (months)	47 ± 32	37 ± 28	NS
Treatment days per patient	2.4 ± 2.6	3.8 ± 2.6	NS
Nr. of indwelling device placements per treatment day	1.0 ± 0.5	0.5 ± 0.4	0.001
Nr. of phlebomoties per treatment day	1.4 ± 1.1	0.5 ± 0.3	0.001
Non-therapeutic level per phlebotomy (%)*	93 ± 16	58 ± 42	0.017

IV UFH: intravenous unfractionated heparin; SC LMWH: subcutaneous low molecular weight heparin; Nr.: number; * activated partial thromboplastin time (PTT) <55 or >85 seconds or anti-Xa level <0,5 or >1,0 U/mL; data shown as meamstandard deviation; p-value: independent t-test.

P1965 - EFFECT OF CARDIAC INTERVENTION ON PARENTING STRESS IN YOUNG CHILDREN WITH CONGENITAL HEART DISEASE IN CENTRAL SOUTH AFRICA THREE MONTH AND SIX MONTH OUTCOMES

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Background: Parents of children with congenital heart disease (CHD) are at increased risk of ongoing stress and psychological problems including anxiety and depression. The parenting stress outcomes of children with CHD in South Africa are unknown. The aim was to determine parenting stress in children with CHD following cardiac intervention.
Materials and Methods: Forty-eight consecutive children, 30 months and younger, and their parents were recruited into this observational descriptive study. Parenting stress was assessed using the Parenting Stress Index- Short Form before cardiac intervention, and at three-month and six-month post cardiac intervention. Parenting stress outcomes were compared over time, and variables associated with parenting stress determined.
Results: Baseline data was collected for 40 parents. Sixty percent of parents (n = 24) experienced clinically significant stress prior to cardiac intervention. Levels of parenting stress declined significantly at both three-month ($p < 0.001$) and six-month post-cardiac intervention ($p < 0.001$). More than 31.8% of parents experienced ongoing stress after their child's cardiac intervention. Neurodevelopmental outcome ($p = 0.03$), perceived health-related quality of life ($p = 0.02$), age at first cardiac intervention ($p = 0.03$) and maternal age ($p = 0.04$) were significantly associated with increased levels of parenting stress.
Conclusion: Most parents experienced clinically significant levels of stress prior to their child undergoing cardiac intervention. Parenting stress declined significantly post-cardiac intervention, but a considerable number of parents experienced ongoing stress. Parents should be screened regularly for risk of psychological problems requiring referral for psychological and educational interventions.

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P2030 - WHAT WILL WE DO TO THE INFECTION

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The totle amount of infected cases in 2014,2015 and 2016 was 78,68,63 respectively in our unit, and the pulmonary infection occupied the biggest part of them. This year one of our main targets is to reduce the infection rate. According to the learning, we have made some measures: 1.To enhance the hands hygiene 2. To shorten the ventilator time 3.To remove tubes after surgery in time. We are going to add more ABHRs,and make a specific staff do the supervision of team members'hands hygiene meanwhile.If the rate is able to be reduced this year, we will do the sharing, making formal procedures for implemention.

P2064 - PAEDIATRIC CARDIAC WARD NURSE LEAD DISCHARGE

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Background: Delay in discharge, on a busy cardiac day unit was noted by staff from parent's complaints to cause unnecessary waits in discharge home in a timely fashion. This did not lead to individualised family centred care. Staff satisfaction around timely discharge was compromised. As a tertiary paediatric cardiac centre travelling times for families' are complex and usually prearranged.
Methodology: Before the programme started there was a review of discharge waiting times. Following a literature review into nurse led discharge, a team training day was arranged to introduce the concept of nurse lead discharge. This was followed by a ward Quality Improvement project. A pilot for 1 month following training and setting standards was used for all suitable patients. Audit forms with times, the patient experience and nursing experience were collated over this period.
Findings: An audit before the implementation of the program showed that in 36 patients the delay in discharge time was 23 hours and 40 minutes with a mean time of 65 minute delay before implementation of nurse led discharge. Post the implementation there were no delays. Feedback demonstrated that the nurse felt empowered by the process. There was greater parental satisfaction as it reduced the volume of discharges occurring at the same time allowing for one-to-one care to the family being discharged.
Discussion: The audit data showed a high number of suitable patients for this nurse led program and with a significant reduction in the length of stay role in the day ward. A follow up call next day was used to safety net the process and for the nurses to evaluate effectiveness of practice.
Conclusion: This audit and change in discharge has been embedded and extended to other patient groups. This has resulted in increased family and staff satisfaction.

P2067 - A NATIONAL QUALITY IMPROVEMENT PROJECT EVALUATING A COMMUNITY BASED EARLY WARNING TOOL FOR INFANTS WITH CONGENITAL HEART DISEASE

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Aim: To evaluate the effectiveness of an Early Warning Tool (EWT) for use by families and Children's Community Nursing teams (CCNT) in making decisions about infants at home and standardizing escalation.
Innovation: Currently there are no CCNT EWTs validated for infants with complex CHD in the community setting or for joint use with parents/carers. The EWT is part of an national vision to develop robust models to support acute care in the community using a virtual ward environment Development and tool design prototype The design is based on current knowledge and clinical experience of CHD using structured key domains, in a traffic light system, as part of the current Home Monitoring educational program. These key domains form part of a weekly clinical assessment of the infant, with local CCNT and telephone consultation.
Evaluation: • Organisation of key domains into mutually agreed visual formatted tool • Focus group work with parents and stakeholders – acceptability to families, understanding of the tool • Trial of the tool multiple sites: o retrospective clinical record review – triggers correctly o scenario based (table top) with stakeholders and clinical experts – correctly triggers with escalation plan o in the clinical setting – correctly triggers with escalation plan.
Implementation into practice: • The EWT would be part of the current Home Monitoring Programme (HMP).
• Structured education and implementation by a dedicated Cardiac Nurse Specialist • Dissemination at a national level with wider application • Parent and stakeholder reported outcome measures.
Other: • Number of triggered events • Deaths and deterioration events would be reviewed as per trust policy and to ensure quality review of the EWT • Close links with Rapid Response meetings at the unexpected death of a infant and the application of the EWT • Survival rates will be monitored and reported at local cardiac meetings and via national data sets (CCAD 2016) • Collaboration with the national children's CHD review.

P2088 - GREAT EXPECTATIONS LOOKING FOR ANSWERS; PARENTS' EXPERIENCE OF CARING FOR A CHILD WITH AN IMPLANTED LOOP RECORDER

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Background: Implantable Loop Recorders (ILRs) are cardiac devices which are increasingly being utilised in the diagnosis and management of unexplained paediatric syncope. Although the efficacy and use of implantable diagnostic devices is increasing, little is known about the impact that ILR's have on the child or family. To date no research has focused on children or parents' experiences; however, anecdotal evidence suggests challenges in adjustment to devices. This qualitative study aims to address gaps in knowledge by exploring parents' 'lived experiences' and concerns in this population.
Method: Semi-structured interviews were conducted with 12 parents (11 mothers, 1 father) of 12 children aged 5-16 years. Interviews were recorded, transcribed verbatim and analysed in

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line with Interpretative Phenomenological methodology. Data analysis was an iterative process bringing the data together into meaningful inductive themes giving rise to super-ordinate themes.
Results: The impact on families' lives were revealed through three main super-ordinate themes: living with uncertainty; disruptive childhood and the impact of technology. Parents reported feeling reassured and safer by the presence of the ILR. However, the longer the device was in place the greater the anxiety. Informing and educating family members, friends, schools and sport coaches on the management of symptoms and device created additional burden. The child's needs were paramount for parents. Pain, body disfigurement, effect on self-esteem and incidence of bullying were reported. On-going issues with equipment failure created disappointment, anger and frustration.
Conclusion: Despite the perceived advantages of ILR, such as ease of implantation and potential for obtaining a diagnosis, the study identified issues which have a negative impact on children and parents. This study provides evidence to target additional supportive measures to families throughout the child's journey, including providing information to schools.

P2449 - FEEDING DIFFICULTIES IN NEONATES FOLLOWING CARDIAC SURGERY DETERMINANTS OF PROLONGED FEEDING TUBE USE

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Background and Hypotheses: For babies with complex congenital heart disease, achieving adequate growth and nutrition can be a challenge. Previous studies show that up to 1 in 2 neonates are not able to completely feed orally after cardiac surgery and are discharged home with a feeding tube. Feeding difficulties in this population are often multifactorial, with pre-, intra-, and post-operative factors influencing a neonate's ability to feed orally. The aims of this study were to examine the prevalence and correlates of feeding difficulties in infants who underwent neonatal cardiac surgery, and to investigate resource utilisation by infants with feeding difficulties.
Materials and Methods: All neonates who underwent their first cardiac surgery at the Heart Centre for Children, The Children's Hospital at Westmead between January and December 2009 were included. Demographic, pre-, intra-, and post-operative data were collected via electronic medical records. For the purpose of this study, feeding difficulty was defined as the requirement for ongoing tube feeding at time of discharge home or transfer to another hospital.
Results: Twenty-four out of a total of 79 neonates (30%) were discharged home or transferred to another hospital with a feeding tube. Feeding difficulties were associated with the presence of a genetic syndrome ($p < 0.0001$), assisted feeding preoperatively (OR = 4.4, $p = 0.03$), and having a palliative procedure prior to biventricular repair (OR = 5.1, $p = 0.02$). Infants with feeding difficulties had significantly more reviews by speech pathologists (M = 5.9, SD = 7.9), dieticians (M = 5.9, SD = 5.4), and cardiac clinical nurse consultants (M = 1.2, SD = 1.4) compared to those without feeding difficulties.
Conclusions: This study identified factors which can be used in the early identification of infants who are at increased risk of feeding difficulties, to help guide the direction of limited health resources, as well as being focal points for future research and clinical practice improvement.

P2451 - COMPARING RETESTING RATES BETWEEN DIFFERENT SCREENING METHODS FOR CRITICAL CONGENITAL HEART DISEASE DATA FROM 112839 NEWBORNS FROM A STATE IN NORTHEAST BRAZIL

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Background: screening for critical congenital heart disease (CCHD) with pulse oximetry is a practical strategy. However, many different protocols exist and little differences between them can lead to a great number of retest rates. This can increase the work of already overloaded nursing teams, mainly in developing countries, and lead to decreased efficacy of the method. So, identifying protocols with lower retest rates is important to an efficient population screening for CCHD.
Material and Methods: data obtained from a large population screening program for CHD in Paraíba – a state from northeast Brazil – was utilized. Only the cases with gestational age of 36 weeks or more and complete data for oxygen saturation from the right arm (RA) and one foot were utilized. A total of 112839 cases were included in this study. Five different protocols were included: oxygen saturation (OS) < 95% in both RA and foot or a difference between them >3% (Jegatheesan). OS <95% in either RA and foot or a difference between them >3% (Fouzas). OS <95% in both RA and foot or a difference between them >2% (Granelli). OS <95% in either RA and foot or a difference between them >2% (Ewer) and a foot saturation <96% (Riede). The Cochran's Q test was utilized, and a p-value 0.05 was considered significant.
Results: the protocol with least retest rates was Fouzas (1.04%) followed by Jegatheesan (1,44%), Ewer (1.83%), Granelli (2.10%) and Riede (5,47%). All protocols showed statistical significance between them.
Conclusion: The Fouzas et al protocol produced the lowest retest rate. This finding is important and helpful to optimize the implementation of new screening politics around the world.

P2453 - NOMOGRAMS FOR PULSE OXYGEN SATURATION FROM A POPULATION SCREENING FOR CONGENITAL HEART DISEASE IN NORTHEAST BRAZIL

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Background: Pulse oxygen saturation (POS) is a great strategy for screening critical congenital heart disease (CCHD). Nomograms

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for pre-ductal and post-ductal POS have been described, but several factors like skin color, ambient temperature, altitude, presence of anemia, and others can modify them. Therefore, the analysis of nomograms in a developing country can be useful to optimize the screening for CCHD.
Material and Methods: data obtained from a screening program for CHD in Paraíba – a state from northeast Brazil – was utilized. Only the cases with gestational age of 32 weeks or more and complete data for oxygen saturation from the right arm (RA) and one foot were utilized. The results from pre-ductal and post-ductal oxygen saturation were compared between them by gender and gestational age. Non-parametric tests (Mann-Whitney) were performed to do so.
Results: a total of 114723 cases were analyzed. The median from pre-ductal and post-ductal saturation was 97% ($p5 = 95\%$ and $p95 = 99\%$). The mean for the difference between saturations was 0.8822% (IC: 0.8763 to 0.8880) and more than 95% of the neonates showed a difference minor or equal to 2%. The pre-ductal and post-ductal saturation was higher in term than preterm neonates ($p < 0.01$). Male patients showed a little, but significant, higher pre-ductal ($p = 0.01$) and post-ductal saturation ($p = 0.03$). However, such differences had no clinical significance.
Conclusion: The nomograms obtained in this study are similar to others obtained in developed countries. Therefore, environment and population differences seem to have no impact on obtained nomograms.

P2494 - DEPRESSION AND ITS INFLUENCING FACTORS AMONG MATHERS OF CHILDREN WITH CONGENITAL HEART DISEASE IN KOREA

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Purpose: Mother of children affected by birth defects suffers from a significant psychological burden across the lifespan, but there have been few studies on this area in Korea. Our purpose was to assess depression among mothers of children with congenital heart disease (MCHD) and to explore factors influencing depression among in Korea.
Method: A total of 182 mothers of affected children aged 0-3 years old and 366 mothers of healthy children (MHC) in the same age took part in the study. Kessler Depression Scale was used to assess maternal depression, and logistic regression models were used to explore the factors influencing depression among MCHD.
Results: Of the mothers, 33.0% reported being mild to moderate depression and 8.5% of the mothers reported severe depression. Also, MCHD were more depressed than MHC. The factors influencing MCHD include; mother's self-efficacy, mother's marital conflict, mother's social support, infant's emotionality, mother's smoking and infant's feeding and eating of daily routine. These variables explained 42.5% of MCHD.
Conclusion: The results of the study suggest that the identified factors for MCHD should be included in an intervention program to reduce the risk of depression.

P2518 - INVESTIGATION AND ANALYSIS OF AIRWAY NURSING IN CHILDREN WITH MECHANICAL VENTILATION

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Objective: To understand the pediatric ICU nurses do airway care for children with endotracheal tubes. To find out problems in clinical airway care and provide some recommendations for clinical improvement, and to help to reduce the incidence of nosocomial respiratory tract infection.
Methods: To observe and record the airway care operations in the pediatric ICU of Shanghai Hospital, and have 60 samples for two hospitals. Do statistical analysis on airway care for child ren and assessment of the situation of children before and after airway care.
Results: Statistical data analysis show that the before and after comparison of Sa02, HR, and airway pressure of children with airway care in two hospital is $P < 0.01$which has highly statistical significance. There are also some common problems such as airway care compliance, appropriate suction, time on airway care and consistency between nursing documentations and orders. Nurses are positive in doing airway care according to children needs. There are 50% nurses can assess children conditions during airway care in B hospital. The respiratory therapist will do chest therapy, monitor SaO2, HR, R, airway pressure which are correlated airway care. Most nurses can follow the standards. But nurses in A hospital can't do assessment well, which will be a danger to nursing safety.
Conclusion: Results showed that reasonable frequency should be necessary. Guidelines should contain the frequency of airway care according to patients conditions like SaO2, HR, sputum. Do documentation and add standards of the depth in suction. Strengthen airway care, nursing assessment and sterile concept of operations. Improve the compliance on standards of hand hygiene, masks and sterile gloves wearing. Put an end to risk factors for nosocomial infections, reduce respiratory tract infections rate and improve quality of care.

P2572 - PRELONGED EXTRACORPOREAL MEMBRANE OXYGENATION FOR INFANT WITH PNEUMOCYST CARNII PNEUMONIA

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The selection of various products of pediatric ECMO oxygenator is impossible for adults. I propose to publish a case of five adults oxygenator replaced for 80 days in an environment that was available for use in ECMO. The patient was admitted to the hospital with fever & cyanosis on the day before admission. He underwent severe acidosis and desaturation and underwent VenoVeno ECMO under severe ARDS and Pneumocyst Carinii Pneumonia diagnosis. In the absence of pediatric ECMO, Maquet PLS was used as the oxygenator and Medtronic 550 was used as the all 1/4 inch line. Method: VenoVeno ECMO was performed by inserting hemocath 14fr into Rt. IJV. To increase the drain flow with desaturation and low flow, Rt. Femoral vein to 8fr. Percutaneous catheter was inserted. We performed open sternotomy at the end of the study because of hemodynamic changes such as decreased blood pressure and hypoxic damage of myocardium. Drain outflow: in RA venous 12fr. Cannular and femoral vein first installed 8fr. Catheter was used as Y shape. Artery inflow: ascending aorta to 8fr. Cannular was inserted and changed to VenoAteriall ECMO. On the third day of ECMO administration, Echo was normal in the finding, and the VV ECMO was determined for longterm management. After insertion of the cannula, it was re-replaced with VV ECMO. Anticoagulation: Nafamostat mesilate 0.3 mg/kg ~ 1 mg/kg(Futhan).Target aPTT 80 sec.

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Most oxygen changes since 3weeks later. During the repeated weaning trial, the increase in TB and hemolysis of the infant were severe, resulting in five PLS oxygen changes. During the treatment, the patient could not use the product suitable for the patient, and there was a large inconvenience such as blood volume loss while using adult oxygenators. However, in a limited medical environment, we will have good results and share it together.

P2649 - TRANSITIONS OF CARE IN CONGENITAL HEART DISEASE VARIABLES ASSOCIATED WITH CLINICAL SETBACKS

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Introduction: Improving outcomes and transitions in care are national initiatives. Hospitalizations in congenital heart disease (CHD) patients are often complex. If variables associated with clinical deterioration can be identified, quality initiatives can prevent "setback" events. In this study, a setback is defined as a clinical deterioration requiring escalation of care within 3 days of transition to step-down unit. The objective was to identify patients at risk for setback and variables associated with changes in clinical status which resulted in return to the cardiothoracic intensive care unit (CTICU). Hypothesis: CHD patients less than 6 months old are at risk for a setback event.
Methods: A retrospective chart review was performed on patients admitted to the CTICU between 01/01/2011 to 12/31/2011 to identify patients transferred to the CTICU within 3 days of transition to the step-down unit. Variables evaluated included diagnosis, age, weight, length of stay, date, time, and reason for setback.
Results: Thirty-seven setback events occurred in 30 patients. Thirteen (35%) were due to post-procedure recovery protocol. In 24 setbacks, occurring in 19 patients, median age was 6.3 months (0-48.7 years); median weight was 6.5 kg (3.1-86.2). Respiratory distress/arrest was the reason for transfer in 19/24 (79%) events with 13/19 (68%) occurring in patients less than 12 months old and 15/19 (79%) in patients less than 8 kg. Four of 19 (21%) patients expired during hospital admission. Median time prior to setback was 30.5 hrs (range 3.9-72.1); 12/24 (50%) events occurred on the weekend; 14/24 (58%) were during off-shift hours.
Conclusions: Children less than 8 kg and less than 12 months are at risk for a setback within 3 days of transfer to step-down unit. Respiratory distress was most frequent clinical reason. Setback events occurred more frequently during off-shift and/or weekends. Further studies of specific variables are needed.

P2705 - IMPROVING EARLY INTERVENTION FOLLOW UP FOR INFANTS WITH CONGENITAL HEART DISEASE

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Background: Infants with single ventricle palliation are at-risk for developmental delays (DD) related to prematurity, cardiopulmonary resuscitation/support, genetic anomalies, and prolonged hospitalizations. Since 2006, our program has provided interstage care for these infants. Through this program, infants are referred to Florida's Early Intervention (EI), which provides developmental evaluations and therapies to children birth to three.
Method: Using our EMR, infants <6 months who underwent a single ventricle palliation were identified. From January 2012 - April 2015, parents received a prescription for an EI evaluation and were instructed to schedule an appointment at four months. Beginning in May 2015, a neurodevelopmental (ND) nurse coordinator directly submitted an EI/therapy referral and followed up to evaluate parenteral compliance.
Results: Eighty infants were identified that underwent a Norwood, shunt placement, ductal stenting, or pulmonary artery banding. From January 2012 -April 2015, 59 patients were identified at-risk for DD and 46/59 patients met criteria for an EI referral (criteria included: EI locally available, patient alive at four months, parent reachable for follow-up). Nineteen/46 patients (41%) complied with a therapy evaluation after discharge. Seventeen/19 (89%) had therapy recommended, and all 17 patients complied with therapies. Between May 2015 - September 2016, using our new approach, 21 patients were identified at-risk and 19/21 patients met criteria for evaluation. Eighteen/19 patients (95%) complied with therapy evaluation after discharge. Sixteen/18 patients (89%) had therapy recommended, and all 16 patients complied with therapies. Three/19 patients (16%) required a referral resubmission because our ND nurse coordinator recognized the therapy site failed to contact the family. *Conclusion:* Single ventricle infants are at-risk for DD. When evaluated by EI these patients qualify and follow-up with therapies. Developmental care improves with a ND nurse coordinator providing direct referral to EI and follow-up to evaluate parenteral compliance.

P2769 - A CALL TO ACTION – IMPROVING THE HEALTH OF THE WORK ENVIRONMENT IN THE CARDIOVASCULAR OPERATING ROOM

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Background/Hypothesis: The American Association of Critical Care Nurses (AACN) has identified systemic behaviors that maintain patient safety, ensure optimal outcomes, and support excellence in nursing practice. These behaviors are organized into six evidence-based standards: skilled communication, true collaboration, effective decision making, appropriate staffing, meaningful recognition and authentic leadership. AACN developed an electronic survey to provide a quantitative assessment of the health of the work environment (HWE). Scoring guidelines are from Needs Improvement, Good, to Excellent. The survey was electronically mailed to Cardiovascular Operating Room (CVOR) interdisciplinary staff from 2010 to 2015. After receiving an overall score within Needs Improvement in 2015, the unit leadership convened to address areas for improvement.
Methods and Materials: A comprehensive and immediate action plan was designed and implemented over 12 months. An authentic commitment to improve the health of the environment was reinforced by cardiovascular nursing and surgical leadership. Based on a key driver diagram, the ambassador group developed and implemented change initiatives and evidence-based practices. Interdisciplinary staff were re-surveyed in 2016.
Results: Eighteen change initiatives and evidence-based practices were implemented to address the areas for improvement.

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The 2016 survey showed an overall improved score as well as improvement within each of the six standards. The overall score and standard scores improved from within Needs Improvement to Good.
Conclusions: Change initiatives were implemented by a nurse-led interdisciplinary team to foster and sustain a commitment to excellence in the work environment. Targeted initiatives will continue to be implemented to address areas requiring improvement. Our results highlight that maintaining and sustaining a healthy work environment is a continuous process and an activity for which all healthcare team members are accountable. Maintaining a culture of health in the work environment will continue to be a priority within CVOR as it is critical to ensuring optimal patient outcomes.

P2772 - TRACHEOSTOMY FOR CHILDREN ADMITTED FOR CONGENITAL HEART SURGERY

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Background/Hypothesis: The objective is to describe the prevalence of tracheostomy placement and outcomes for children during admission for congenital heart surgery (2006,2009).
Methods and Materials: ICD-9-CM codes were used to identify patients <18 years of age undergoing both congenital heart surgery and tracheostomy during admission using Health Care Utilization Project Kids Inpatient Database (KID) year 2006 and 2009. Hospital characteristics and patient demographic, clinical and surgical (RACHS-1 risk categories) characteristics, discharge disposition, and inpatient utilization were summarized.
Results: In 2006, 16,891 cases were admitted for congenital heart surgery of which 179 (1.1%) also underwent tracheostomy. In 2009, 18,765 cases were admitted of which 234 (1.2%) underwent tracheostomy. In both years, the rate of tracheostomy varied significantly by region, with the highest percentage observed in the South (33.5% in 2006; 33.5% in 2009). Among those who had both procedures, the rate of in-hospital death decreased from 2006 to 2009 (26.8% and 16.2%, respectively). When comparing 2006 to 2009, the use of Medicaid insurance decreased (64.8% to 58.6) and use of private insurance increased (29.6% to 35.5%). The percentage of patients with prematurity, non-cardiac anomalies, or non-Downs major chromosomal abnormalities increased. In 2006 and 2009, the majority were categorized as RACHS-1 categories 2, 3, and 4 (56% and 38.5%, respectively). The percent of cases categorized as a RACHS-1 category 6 increased from 2006 to 2009 (1.7% to 4.7%). The median length of stay was 132 days (16 to 356) in 2006 and 139 days (12 to 362) in 2009. In 2006, 38% were discharged from a children's hospital. In 2009, 31.6% were discharged from a children's hospital.
Conclusions: Tracheostomy, although rare during congenital heart surgery admission, is associated with high in-hospital mortality and resource use. Despite having a similar rate of tracheostomy and RACHS-1 classifications, the rate of in-hospital mortality decreased by 10.6%.

P2777 - MANAGING CARDIAC INTENSIVE CARE UNIT CAPACITY STRATEGIC SOLUTIONS

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Background/Hypothesis: For an extended period of time during summer of 2014, the needs of our cardiac patients exceeded the capacity of our physical Cardiac Intensive Care Unit (CICU). In response to this unprecedented demand on resources, an interdisciplinary group was established to develop strategic solutions for CICU capacity management.
Methods and Materials: The interdisciplinary group met regularly with an external consultant to analyze the capacity impact of patients remaining in the CICU for more than 28 days. Using this cohort of 182 long-stay patients, we developed a model to predict ICU length of stay to inform scheduling of cardiac surgical cases. A daily data dashboard was generated to display the daily census, incoming cardiac catheterization and surgical cases, and a prediction of the weekly capacity. In addition, a smaller interdisciplinary admissions group was developed to assess CICU capacity daily and advise on pending admissions and transfers. A brief daily huddle was implemented for the admissions group to review the predictions and dashboard to ensure adequate capacity management. At six months, these strategic solutions were evaluated to determine feasibility and sustainability.
Results: Since May 2015, a brief daily huddle of the admissions group has been held every morning. During each huddle, the predictions from the model and the daily dashboard are used to inform surgical scheduling as well admission and transfer decisions. It has been feasible to update and use the data dashboard and predictions daily. During the summer of 2015, there were zero cardiac surgical patients cancelled or postponed due to lack of CICU bed availability.
Conclusions: The development and implementation of strategic solutions for capacity management in the CICU has resulted in a dramatic reduction in the number of cardiac surgery cancellations or rescheduling. The strategic solutions have been found to be feasible to implement and sustain.

P2799 - COMPREHENSIVE MEASUREMENT OF NURSING PRACTICE IN THE CARDIAC INTENSIVE CARE UNIT USING THE CAMEO

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Background/Hypothesis: Efforts to capture nursing workload have focused on describing and quantifying direct care nursing activities. Existing tools do not account for many of the provider-related "invisible" activities that comprise critical thinking, leading to an incomplete assessment of the cognitive workload. Complexity Assessment and Monitoring to Ensure Optimal Outcomes (CAMEO) is a validated instrument that provides a quantitative measurement of cognitive workload required to deliver pediatric nursing care. It includes direct and indirect nursing care activities and incorporates the intellectual processing of data and critical thinking

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necessary to deliver high-quality, holistic care to patients and families. Complexity is measured using a classification scoring system with I representing stable patients requiring minimal intervention and V representing patients with clinical instability and/or complex care coordination needs. The objective was to assess the contribution of indirect nursing care in the cardiac intensive care unit (CICU).
Methods and Materials: Completed CAMEOs were categorized and summarized by level of complexity. Reported workload for the domains of care coordination, family assessment and support, and education and discharge teaching were calculated by number of reported actions and time required to complete the activity by the CICU nurse.
Results: Among 539 CAMEOs reviewed, 90.4% were categorized as classification III, IV, or V. The number of indirect care activities increased with level of complexity with an average of 3.38 care activities reported for level V. Among three indirect care domains, the most frequently reported activity was identified as care coordination. As level of complexity increased so did the amount of time spent to support these indirect activities with 50.5% of level V care reporting >30 minutes.
Conclusions: To understand true workload of pediatric critical care nursing, both direct and indirect care must be considered. Use of CAMEO for pediatric cardiovascular patients revealed a consistently complex level of nursing care.

P2805 - BALANCING AUTONOMY AND INVOLVEMENT THE ROLE OF THE FATHER IN DECISION MAKING FOLLOWING DIAGNOSIS OF A SEVERE FETAL ANOMALY A QUALITATIVE STUDY

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Background: As the discourse surrounding 'new-fatherhood' has become established in the literature, the experiences of fathers-to-be has increasingly been reported. However, the role of fathers-to-be in the decision-making process following diagnosis of a severe fetal anomaly has predominantly been overlooked. Aims: Drawing on findings from a larger study, this article begins to fill that gap by exploring tensions arising from prioritising maternal identity over paternal/couple identities, and seeks to identify mechanisms through which one can be supported without disempowering the other.
Methods: Data from interviews with parents-to-be and clinicians, and audio-recorded consultations were collated. Analysis was undertaken using a constant comparative based approach.
Findings: Data collated from the fathers' interviews highlighted a sense of being a 'bystander', a role that resonates with men's experience of pregnancy/childbirth more broadly. Whilst maternal identity is reinforced from the physical changes in her body to the legal requirements of her signature on consent forms, the paternal identity exists only when validated by others. "Disenfranchised grief", a grief that is not or cannot be openly acknowledged, was frequently encountered by fathers-to-be. This is reflected in the social expectations of stoicism and hidden grief. Implications for practice: Symbolic procedures can be implemented to support the couple as an entity without disempowering the mother. Examples such as providing opportunities for both parents-to-be to sign consent forms when a termination has been decided upon, engagement of the father-to-be by documenting his name in fetal medicine notes so healthcare professionals can refer to him by name, and practical aspects such as ensuring both parents have access to basic facilities such as bedding and food whilst in hospital. As the 'patient', these items will automatically be provided for the mother, but as a 'visitor' the father, as revealed in many of the experiences recalled in this study, was frequently overlooked.

P2828 - READINESS FOR TRANSFER IN ADOLESCENTS WITH CONGENITAL HEART DISEASE

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Introduction: Many patients with congenital heart disease (CHD) are in need of lifelong medical follow-up. Transfer to adult care in most cases is determined by the age of the patient. In order to develop interventions that can help the young person in this transfer, it is important to determine their readiness to transfer to this new setting.
Aim: The aims were (i) to describe the level of readiness for transfer in adolescents with CHD, and (ii) to explore the association of transition readiness with age, sex and disease complexity.
Method: In a cross-sectional study, 203 adolescents from four outpatient paediatric cardiology units in Sweden participated (response rate: 34%). The mean age was 15.7 ± 1.2 years and 45% were females. Patients could be included if they met the following criteria: 1) diagnosed with CHD; 2) aged 14-18 years; and 3) under active follow-up at one of the participating centres. The overall transition readiness was measured using the Readiness for Transition Questionnaire (RTQ). The score ranges from 2-8, with higher scores denoting higher level of readiness.
Results: Mean RTQ-score in 14 year olds was 4.6 ± 1.6 ($n = 37$), in 15 year olds 5.1 ± 1.8 ($n = 51$), in 16 year olds 5.2 ± 1.6 ($n = 57$), and in 17 year olds 5.9 ± 1.8 ($n = 68$). Indeed, older participants had a significantly higher RTQ-score ($p = 0.003$). Male patients showed to have a higher RTQ mean score (5.5 ± 1.7) than females (5.0 ± 1.8)($p = 0.05$). Transfer readiness was not associated with sex or complexity of the condition (mild 5.3 ± 1.6; moderate 5.3 ± 1.9; severe 5.2 ± 1.8; $p = 0.98$).
Conclusion: Self-reported overall RTQ increased with age. This information is relevant when setting up intervention studies. There is a natural growth in readiness regardless the complexity of the CHD. Interventions should be able to outstrip the natural evolution. Future studies should further explore transition readiness in terms of patients' and parents' involvement and responsibility of care.

P2850 - SKIN PROTECTION POLICY FOR CRITICAL PEDIATRIC PATIENTS WITH CONGENITAL HEALTH DISEASE HAVING EXTRACORPOREAL MEMBRANE OXYGENATION (ECMO) THERAPY A SUCCESSFUL EXPERIENCE LEAD BY NURSES

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Background: Since 2015 The Fundación Cardioinfantil Instituto de Cardiología (FCI-IC) in Bogotá, Colombia implemented the skin protection for all children having extracorporeal membrane oxygenation (ECMO) therapy in the cardiovascular intensive care unit (CICU). The implementation of preventive care strategies was lead by the nurses of the Skin Care Program (SCP) and from the CICU.
Objective: to describe (1) the nursing process of skin care assessment, administration of protective skin strategies, evaluation and following up of outcomes during hospitalization, (2) the characteristics of the pediatric population under care, and (3) the effectiveness of the program in the prevention of pressure ulcers.
Methods: All pediatric patients (2015-2016) with congenital heart disease eligible for ECMO received the standardized skin protection. Parents of patients signed a consent form for care implementation. Each patient was assessed and intrinsic and extrinsic factors were taking into a model a care. The skin areas with high risk were protected with hydrocolloid dressings and other areas were protected with hyper-oxygenated fatty acid esters. Following up of the patients was daily by CICU nurses and each five days by SCP nurses.
Results: Nine children from a total of 1204 admitted to the CICU, with mean age of 9.3 months, six men and three women were included in the cohort of patients having ECMO at the FCI-IC. The most prevalent complication was ventricular dysfunction after surgery. The mean length of stay was of 31 days and the mean time in ECMO was 13 days. There were three (0.3%) pressure injuries (Grade I) in occipital area with epithelialization in 4-5 days. Patients did not have pressure ulcers compared with patients not having ECMO (ulcer rate of 6.6 ×1000).
Conclusion: This study highlights the benefit of the implementation of one standardized policy of skin preventive protection for pediatric patients under high risk.

P2864 - PREPLANNED AND PREPREPARED DISCHARGE RAISED QUALITY

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The cardiac nurse practitioner team took on the management of discharges from the cardiology ward environment in 2011. The flow of patients in any large tertiary cardiac paediatric centre is crucial. This will ensure safety, quality of care and efficiency. Discharge of a patient with agreed clinical safety and who continues to receive high quality and efficiency of care is crucial to patient flow and the ability to ensure more patients access the service. This process is crucial for admission of the next patient requiring planned or emergency cardiac surgery, as part of daily planning strategy.
Method: The unit currently runs at over 87% capacity, which means that each bed day is very precious to the hospital and to the patient. An understanding of the discharge planning is critical to running a service where admissions are as critical as your discharges. The collaborative working of clinical experts is very important and the expertise gained working in this "specialist area " or practice cannot be underestimated. Family expectations and preparation were undertaken as part of a Nurse Led clinic to reward the individual and family. Patient supported outcomes measures were used to measure success. With this development it was obvious from data collection that their were large gaps in service and therefore team developments and wins e.g. assessments, wound reviews, follow up calls and animal reviews.
Results: Data collected shows the development of this service and the ANP role. Data will be presented to support this development. Family reviews will provide balancing measures. The success of this model with a quality improvement connection, has been used in other areas of clinical care.
Outcomes: Outcomes measures are needed to ensure the qualitative and quantitative measures are used to support future developments.

P2881 - PERCEIVED CARDIAC CONSEQUENCES FROM ADOLESCENTS WITH TETRALOGY OF FALLOT AND REVIEW OF CLINICAL DATA

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Introduction: Tetralogy of Fallot (TOF) may have long term consequences with exercise ability, quality of life and need for future cardiac surgery. Adolescents may have perceptions of these consequences which may or may not be valid.
Methods: Adolescents with TOF were asked to completed questionnaires to describe how their congenital heart disease affected their exercise abilities, and to report their perceptions of the need for future surgery and long term consequences. Reported symptomatology and quantitative data obtained from echocardiograms, electrocardiograms (ECG), cardiac stress tests, and cardiac MRIs were reviewed and compared to the responses.
Results: Forty percent of TOF adolescents reported no long term cardiac consequences or effect on their exercise abilities. Exercise limits were perceived with minimal consequence in 37% and moderate consequences in 13%. Despite rarely applied exercise restrictions from providers, 28% of adolescents perceived an exercise restriction was recommended. The most commonly reported complaints were exercise intolerance and fatigue. The majority of echocardiograms revealed normal ventricular function, minimal pulmonary valve stenosis; half revealed moderate right ventricle dilatation and half revealed aortic root enlargement. Nearly 50% of patients, completed Holters, exercise stress test and/or cardiac MRI. No significant rhythm abnormalities were seen on the cardiac tracings. Premature ventricular complexes were infrequently reported. Cardiac MRIs provided qualitative data on the pulmonary valve insufficiency. Possible or more definite need for surgery was reported in 70%. Adolescents reporting greater limitations or concerns for future surgery were more likely to have completed a Holter or MRI with more significant cardiac findings noted.
Conclusions: The quantitative data from the cardiac tests frequently validated the adolescents perceived greater concerns with exercise abilities, need for future surgery or long term consequences.

P3015 - CLINICAL HAND OFF IMPROVEMENT INITIATIVE TO STANDARDIZE INTERDISCIPLINARY PATIENT HAND OFF

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Objective: The Clinical Handoff Improvement (CHAIN) initiative aimed to develop and implement a highly reliable and sustainable

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standardized patient handoff process across all care areas in the Heart Center to optimize the transfer of accurate information and ensure against handoff-related care failures.
Methods: An interdisciplinary quality improvement team was formed to improve patient handoff. Improvement was guided by evidence-based literature, the hospital's clinical handoff policy and mission, and contributions from clinicians across the Heart Center continuum. The CHAIN working group developed an I-PASS (illness severity, patient summary, action list, situation awareness and contingency planning, synthesis by receiver) tool specific to the cardiac units and handoff process involving team-based face-to-face communication. The initiative was piloted over two months for patient transfers from the Cardiac Intensive Care Unit (CICU) to the Cardiac Inpatient Unit. Phase two included scheduled times for structured handoffs. Evaluation via direct observation and surveys included measuring team compliance of the I-PASS format, duration and efficiency of the handoff, synthesis by the receiving team, and clinician satisfaction.
Results: From August to September 2016, 26 patient handoffs were observed (88% from surgical service and 12% from medical service). Duration of team-based handoff decreased with Phase 2 of the pilot (average of 8 minutes per handoff). Overall, clinician satisfaction from both sending and receiving teams improved with the implementation of scheduled times.
Conclusions: Implementation of a structured handoff initiative was found to be both feasible and successful. The CHAIN working group was able to use the pilot to analyze content and structure of handoff communication. Since the pilot, the standardized team-based patient handoff has been implemented for all handoffs between the CICU and inpatient unit. The pilot provided insight to expand the team-based process to handoffs across the Heart Center.

P3019 - CONCERNING THE CHEWS STAFF AND FAMILY CONCERN CONTRIBUTE TO THE IDENTIFICATION OF HOSPITALIZED CHILDREN AT RISK FOR CRITICAL DETERIORATION

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Background: Early warning scores are used in hospitals to identify patients at risk for critical deterioration. The Children's Hospital Early Warning Score (CHEWS) is the only pediatric early warning score that includes clinician and family concern into the aggregate score. The objective was to determine the validity of 'staff concern' and 'family concern' within CHEWS at identifying children at risk for critical deterioration.
Methods: A retrospective cohort study was used to examine the association between 'staff concern' or 'family concern' and critical deterioration in pediatric cardiac (n = 312) and non-cardiac (n = 1136) inpatients. Odds ratios (OR), sensitivity, specificity, and the area under the receiver operator characteristic curve (ROC) were estimated for the CHEWS domains.
Results: See Table 1 and 2.
Conclusion: 'Staff concern' had the highest discrimination at identifying pediatric critical deterioration. The presence of 'staff concern' and 'family concern' were both statistically significant in patients that experienced critical deterioration compared to control patients. The OR for critical deterioration was three times higher in the cardiac patients compared to the non-cardiac patients when staff concern was present. Staff concern' and 'family concern' are valid CHEWS domains in the identification of hospitalized children at risk for critical deterioration.

Table 1. Odds ratio (OR) for critical deterioration by CHEWS domain.

	Non-Cardiac Sample		Cardiac Sample	
CHEWS Domain	**OR**	**p value**	**OR**	**p value**
Staff Concern	10.37	<0.001	31.4	<0.001
Family Concerned or absent	2.87	<0.001	0.96	0.96
Behavior/Neuro	1.96	<0.001	1.7	0.057
Cardiovascular	1.24	0.02	2.25	<0.001
Respiratory	1.64	<0.001	2.47	<0.001

Table 2. Area under the receiver operating curve (ROC), sensitivity, and specificity of 'Staff Concern' and 'Family Concern or Absent' CHEWS domains.

	Non-Cardiac Patients	Cardiac Patients
Staff Concern		
ROC	0.856	0.890
Sensitivity	83.1%	96.2%
Specificity	88.2%	91.7%
Family Concern or Absent		
ROC	0.766	0.661
Sensitivity	78.1%	64.1%
Specificity	75.2%	68.0%

P3023 - POST ORIENTATION SUPPORT TARGETING EDUCATION AND RESILIENCE PROGRAM (POSTER) FOR NEWLY HIRED NURSES ON AN ACUTE CARE PEDIATRIC CARDIOLOGY UNIT

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Background: New nurses may have high levels of psychological distress, low resiliency, and role transition challenges resulting in decreased confidence, marginalization, and potential errors. Satisfaction among new nurses improved if they had access to supportive resources and were aware of professional development opportunities. New nurses reported higher levels of security off orientation when they received proactive support including regular check-ins, time set aside for them, and monitoring of their progress.
Methods: the Inpatient Cardiology Education Team initiated the Post-Orientation Support Targeting Education & Resilience (POSTER) Program to support new nurses. All new nurses were enrolled in the POSTER program upon orientation completion. The new nurse's preceptor completes an aggregate POSTER scale (0–36) comprised of clinical practice, education needs, psychosocial support and experience level to identify the level of needed support. The POSTER program provides twice a day structured 'check-in's with the new nurse for 6-12 months depending on need. Each new nurse is matched to a nurse educator as their personal POSTER coach. Coaching sessions are provided initially on a monthly basis and decreased if indicated. The new nurse 'graduates' from the POSTER program into the unit's peer-based mentoring program after mutual agreement with the POSTER coach.
Results: 42 nurses entered the POSTER program in 19 months, less than half (38%) were new graduate nurses. Retention of the

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nurses was 88% (n = 37) for the unit and 95% for the department. Initial POSTER scores ranged from 1-21 with a mean of 7.0, 6.5 for unit-retained nurses and 17 for those who left (n = 5) (p = 0.0001).
Conclusion: The POSTER program had a high retention rate since implementation. The POSTER scale may be helpful in identifying nurses completing orientation that are at risk for leaving their position.

P3045 - IMPROVING COMPLIANCE WITH "SAFE SURGERY CHECKLIST" PROCESS; A MULTIDISIPLINARY IMPROVEMENT PROJECT

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In this study we aimed to improve compliance with Safe Surgery Checklist (WHO) to reduce surgery related complications. Compliance with the checklist in June 2014 was 57%. A multidisciplinary group composed of frontline staff gathered to define the root causes of non-compliance. Individual interviews were also used to gather information. The main root causes were: the documentation process had both manual and electronic phases leading to confusion, the checklists had many questions some of which had no value, leading to frustration and time loss, the use of electronic checklists prevented the optimal practice of time out facing the patient. The group decided on the following interventions: Simplification of the forms and getting rid of the wastes, multiple forms were combined to reduce handoff problems and improve communication, moving to manual forms to prevent variation and establish bedside time out. The interventions led dramatic improvement in compliance from 57% in 2014 to 88% in 2015. The next step was to deep dive into specifics where we recognized that the department of cardiovascular surgery (CVS) needed department specific attention. We conducted education sessions specific to CVS to tackle specific concerns and discuss them. Meanwhile, the Anesthesiologists were given the right not to put patients into sleep until proper Time out was performed. These actions increased compliance of cardiovascular surgeons from 79% to 97%. The overall improvement was sustained at the level of 88% in 2016 by regular reporting and personal feedback to non-compliant practitioners. Conclusion: a multidisciplinary team is essential for defining the root causes and actions for non-compliance. A user-friendly documentation which provides the chance to communicate with the patient and team improves compliance with safe surgery checklist. Working with frontline staff is an essential token to improve compliance. A powerful leadership support is also essential to implement and sustain improvement.

RHEUMATIC

P1100 - ECHOCARDIOGRAPHIC PREVALENCE OF RHEUMATIC HEART DISEASE IN 4515 SUDANESE SCHOLARS EVIDENCE OF HEALTH INEQUITY

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Introduction: Rheumatic heart disease is the commonest cause of acquired heart disease in Sudan with a clinical prevalence of 11 per 1000 in 1992.. Introduction of hand held echocardiography (HHE) enabled the detection of subclinical disease which was found to be several folds higher than clinical prevalence in many countries. We carried this study to measure the echo prevalence and initiate a prevention program.
Methodology: This is a prospective epidemiological study on scholars 10-15 years of age. Phase 1 was conducted in Khartoum (September 2015- February 2016). Phase 2: Conducted in South Darfur, Niyala camps for displaced people (July - September 2016). Echocardiographic (echo) screening using HHE (V scan-GE) was conducted In Khartoum by pediatric cardiology fellows using the World Heart Federation criteria. While in Niyala, a simplified protocol using one echo view performed by trained medical officers was utilized. Suspected cases underwent detailed echo examination using standard echo (SE) machine. Training of health personnel was conducted through lectures and workshops.
Results: In Khartoum a total of 3000 scholars were screened, 8 cases were found to be positive for RHD by HHE. Out of them, only 1 case was confirmed by SE. The echo prevalence of RHD was 0.3 per 1000.In Darfur, a total of 1515 cases were screened. By HHE, 59 cases were positive for RHD. Out of these 34 underwent SE; 29 of whom were found to have RHD; 22 had definite and 7 had borderline disease. HHE sensitivity was 85.2%. The estimated prevalence was 30.7 per 1000. A total of 779 health worker were trained and 50 000 posters and pamphlets were distributed. The striking difference of RHD prevalence in the 2 areas is an indication of health service inequity and the need to mobilize resources to primary health care in remote areas.

P1108 - EPIDEMIOLOGY OF CONGESTIVE HEART FAILURE IN CHILDREN IN A TERTIAIRY AFRICAN CENTRE

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Background: The aim of the study was to investigate the epidemiology of congestive heart failure in children in St. Elizabeth Catholic General hospital, cardiac centre.
Patients and Methods: This retrospective analysis included 265 patients aged between 5 and 16 years old who consulted in the Cardiac centre from July 2008 through July 2013. Data from patients' records, two-dimensional echocardiographic studies, electrocardiogram were reviewed.
Results: Patients aged between 4 days and 16 years old with a mean age of 8,4 ± 4,5 years. The main symptoms were: tachycardia, growth retardation, cyanosis, cough in patients with congenital heart diseases, shortness of breath, lower limbs and abdominal swelling, orthopnoea in patients with acquired heart diseases. Acquired heart diseases due to post rheumatic valvulopathies were mostly diagnosed in 190 cases (71%), severe mitral regurgitation being the commonest diagnosis. Less were mitral stenosis and combined pathologies (mitral and aortic valves) and HIV cardiomyopathy. In the group of patients with congenital heart diseases, ventricular septal defect defect was the commonest diagnosis. Tetralogy of Fallot, patent arterial duct, atrial septal defect, coartation of the aorta, complex congenital heart pathologies and sickle cell cardiomyopathy were also seen. Corrective and palliative surgery were performed in patients with indication after stabilization.

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Conclusion: Post rheumatic mitral valve regurgitation is the pathology the most encountered in children with congestive heart failure having acquired cardiopathies and ventricular septal defect is the pathology the most encountered in children with congestive heart failure having congenital cardiopathies. Due to financial limitation poverty and illiteracy of parents, the follow up of patients is difficult.

P1236 - ADHERENCE TO SECONDARY ANTIBIOTIC PROPHYLAXIS FOR PATIENTS WITH SCREENING DETECTED RHEUMATIC HEART DISEASE

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Background: Echocardiographic screening for rheumatic heart disease (RHD) can detect subclinical cases, however adequate adherence to secondary antibiotic prophylaxis (SAP) is required to alter disease outcomes. We aimed to investigate the adherence to SAP among young people with RHD diagnosed through echocardiographic screening in Fiji, and to investigate factors associated with adherence.
Methods: Patients diagnosed with RHD through echocardiographic screening in Fiji from 2006 – 2014 were included. Dates of benzathine penicillin G injections were collected from 76 health clinics nationally from December 2011 to December 2014. Adherence was measured using the proportion of days covered (PDC). Multivariate logistic regression analysis was used to identify characteristics associated with any adherence (≥1 injection received) and adequate adherence (PDC ≥ 0.80.
Results: Of 494 patients, 268 (54%) were female and the median age was 14 years. Overall, 203 (41%) had no injections recorded and just 33 (7%) had adequate adherence. Multivariate logistic regression showed increasing age (OR 0.93 per year, 95% CI 0.87-0.99), and time since diagnosis ≥1.5 years (OR 0.53, 95% CI 0.37–0.79) to be inversely associated with any adherence. Non-iTaukei ethnicity (OR 2.58, 95% CI 1.04-6.33) and urban residence (OR 3.36, 95% CI 1.54–7.36) were associated with adequate adherence, whereas time since diagnosis ≥1.5 years (OR 0.38, 95% CI 0.17–0.83) was inversely associated with adequate adherence.
Conclusions: This is the first study to assess adherence to SAP following echocardiographic screening, and the first to utilise the PDC as an adherence measure for RHD. Adherence in Fiji is currently inadequate for individual patient protection or population disease control. Secondary prevention should be strengthened before further screening can be justified.

P1266 - ECHOCARDIOGRAPHIC SCREENING FOR RHEUMATIC HEART DISEASE (RHD) AMONG ASYMPTOMATIC ADULTS 18 25 YEARS OF AGE STUDY FROM A REGION WITH HIGH PREVALENCE OF ECHOCARDIOGRAPHIC RHD IN ASYMPTOMATIC SCHOOL CHILDREN

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Background: Several studies, including our own, have reported a high prevalence of rheumatic heart disease (RHD) in school children on echocardiographic screening. However the significance of these echocardiographic abnormalities remains ill defined. A long term follow up of such children will be ideal to understand the natural history of mild, subclinical RHD. Screening adults living in the same region may provide some insight into the natural history.
Methods: We carried out a cross sectional survey among asymptomatic adults 18-25 years of age in a pre identified area of northern part of India where an echocardiographic screening among school children had detected a high prevalence of RHD. The demographic data was collected. After a brief history and physical examination, echo Doppler was performed by an experienced cardiologist. A diagnosis of RHD was made using the World Heart Federation criteria (WHF); it was divided into "definite RHD" and "borderline RHD".
Results: A total of 835 adults have been screened over the preceding 6 months. The mean age was 20.6 ± 2.5 years, 406 (48.6%) were males. Clinical examination detected RHD in three cases (clinical RHD: 3.6/1000). Using the WHF criteria, RHD was diagnosed in a total of 13 cases (15.6/1000; 95% CI 9.1-26.4/1000), 5 were diagnosed as definite RHD and 8 as borderline RHD. RHD was significantly more common in females and in families with more overcrowding in the house ($p < 0.01$ for both).
Conclusions: This preliminary work demonstrates that prevalence of RHD is high among asymptomatic adults in this region of northern part of India where an earlier study had shown high prevalence in school children. This data may be useful to understand the natural history of mild, subclinical RHD since it may act as a surrogate marker for long term follow up of children diagnosed to have RHD on screening echocardiography.

P1277 - ECHOCARDIOGRAPHIC PROGRESSION OF SCREENING DETECTED DEFINITE AND BORDERLINE RHEUMATIC HEART DISEASE

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Background: Echocardiography is a sensitive test for rheumatic heart disease (RHD) screening, however the natural history of RHD detected on screening has not been established. We aimed to evaluate the progression of screening-detected RHD in Fiji.
Methods: All young people previously diagnosed with RHD through screening, with echocardiograms available for review, were eligible. All baseline echocardiograms were re-reported. Participants underwent follow-up echocardiography. A paediatric cardiologist determined the diagnosis using the World Heart Federation criteria and assessed the severity of regurgitation and stenosis.
Results: Ninety-eight participants were recruited (mean age, 17 years; median duration of follow-up, 7.5 years). Two other children had died from severe RHD. Fourteen of 20 (70%) definite RHD cases persisted or progressed, including four (20%) requiring valve surgery. Four (20%) definite RHD cases improved to borderline RHD and two (10%) to normal. Four of 17 (24%)

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borderline cases progressed to definite RHD (moderate: 2; severe: 2) and two (12%) improved to normal. Four of the 55 cases reclassified as normal at baseline progressed to borderline RHD. Cases with a follow-up interval greater than five years were more likely to improve (37% vs 6%, $p = 0.03$).
Conclusions: This is the longest follow-up study of screening-detected RHD and demonstrates the natural history is not benign. Most definite RHD cases persist and others may require surgery or succumb. Progression of some borderline cases to severe RHD demonstrates the need for monitoring and individualised consideration of prophylaxis. Robust health system structures are needed for follow-up and delivery of secondary prophylaxis if RHD screening is to be scaled-up.

P1278 - CLINICAL OUTCOMES FOR SCREENING DETECTED AND CLINICALLY DIAGNOSED RHEUMATIC HEART DISEASE PATIENTS

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Background: Population-based echocardiographic screening is being considered as a disease control strategy for rheumatic heart disease (RHD) in several countries. However, assessment of the utility of screening is limited by the paucity of data on the clinical outcomes of young people with screening-detected RHD. We aimed to describe the outcomes for a cohort with screening-detected RHD, in comparison to patients with clinically-diagnosed RHD.
Methods: A retrospective cohort study included all young people with screening-detected RHD in the Central Division of Fiji in the primary cohort. Screen-negative and clinically-diagnosed comparison groups were matched 1:1 to the primary cohort for baseline demographic characteristics. Data were collected on mortality, clinical complications and healthcare utilisation from the electronic and paper health records and existing databases. Incident Rate Ratios (IRR) and Kaplan-Meier survival curves were used to compare groups.
Results: Seventy participants were included in each group. Demographic characteristics of the groups were very similar (mean age 11 years, 69% female, median follow-up 7 years). There were nine (13%) RHD-related deaths in the clinically-diagnosed group, one (1%) in the screening-detected group (IRR: 9.6, 95% CI 1.3–420.6) and none in the screen-negative group. Complications of RHD were observed in 39 (56%) clinically-diagnosed cases, four (20%) screening-detected cases and one (1%) screen-negative case. There were significant differences in the complication-free survival curves of the groups ($p < 0.001$). Rates of admission and surgery were highest in the clinically-diagnosed group, and higher in the screening-detected than screen-negative group.
Conclusions: This study provides the first evidence that young people with screening-detected RHD have worse health outcomes than the screen-negative population. The prognosis of clinically-diagnosed RHD in Fiji remains very poor, with very high rates of mortality and complications. These data can inform development of evidence-based screening policy. Implementation of comprehensive RHD control strategies are urgently required for prevention of disease and premature mortality.

P1324 - LONG TERM FOLLOW UP STUDY OF PATIENTS WITH KAWASAKI DISEASE

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Background: Kawasaki disease (KD) may result in coronary aneurysm formation and increased risk of cardiovascular complications such as ischemic heart disease. Therefore, the early detection, non-invasive monitoring and long-term follow-up of myocardial ischemia are essential. Purpose of this study is to investigate the long-term prognosis of patients with Kawasaki disease in Korea, and discuss the need for long-term follow-up.
Methods: The subjects were 48 patients among 354 who had been hospitalized due to Kawasaki disease, and who consented to echocardiography and exercise challenge testing. The mean duration from the onset of disease to follow-up testing after rehospitalization was 11.6 years (8.2–17.0). Patients without coronary artery aneurysms at the initial presentation of the disease were classified in group 1, and patients with small aneurysms were in group 2. Test abnormalities and differences between the two groups were analyzed.
Result: There were no significant differences in the results of follow-up echocardiography and exercise challenge testing between the two groups. Although no abnormal findings were noted at follow-up in most patients, a 9-year-old boy in group 2 showed coronary artery dilation. The exercise test indicated normal results in both groups, and echocardiography results were also normal in 100% of cases in group 1 and 93.3% of cases in group 2.
Conclusion: As some patients with coronary aneurysms showed coronary artery dilation, we believe that long-term follow-up may be selectively required in patients with coronary artery complications.

P1325 - ECHOCARDIOGRAPHIC SCREENING OF 3 901 NIGERIAN SCHOOL CHILDREN FOR RHEUMATIC HEART DISEASE PRELIMINARY REPORT

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Background: Echocardiographic screening for Rheumatic Heart Disease (RHD) in Africa has revealed prevalence rates in the range of 0.5–3.9%.
Objective: To determine prevalence of RHD in a large population of Nigerian school children using echocardiography.
Methods: Using portable echocardiography and auscultation, 3,901 school children aged 5years to 16years in Lagos, Nigeria were screened for RHD. Multi-stage sampling of public schools within and outside Lagos metropolis was done to enroll 20 schools. Diagnosis of RHD was based on the 2012 World Heart Federation echocardiographic criteria.

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Results: The 3,901 children screened had mean age of 11.3 ± 2.7 years; 2,085 (53.4%) were females. The majority of them belonged to lower socioeconomic class. There were 35 abnormal echocardiograms. Eight (0.2%) had RHD including 2 cases of definite RHD and 6 cases of borderline RHD giving a prevalence of 2.1/1000 [2.6/1000 in the peri-urban, 1.5/1000 in the urban area). The majority (88%) of children with RHD had mitral valve affectation. Six of the eight children with RHD were males including all the children with definite RHD. Mean age of children with and without RHD were similar (12.1 ± 2.1 years versus 11.3 ± 2.7 years respectively; p = 0.4) Echocardiography detected RHD 4 times better than auscultation [echocardiography 8 (0.2%) versus auscultation 2 (0.05%); p < 0.0001]. All 8 cases of RHD were asymptomatic except for heart murmur in the 2 definite cases. The remaining 27 cases with abnormal echocardiograms had congenital heart diseases.
Conclusion: Prevalence of RHD among school children in Lagos, Nigeria is low compared to other African countries. Mitral valve is the most commonly affected valve. All children with RHD were asymptomatic. To detect one case of RHD among apparently healthy school children in Lagos, Nigeria, 400 children have to undergo echocardiography.

P1340 - MULTI CENTERAL STUDY ON IVIG TREATMENT TO KAWASAKI DISEASE

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Objective: To investigate treatment in children with Kawasaki disease by three different dose of IVIG therapies and to seek for a most effective therapy.
Methods: We did a multicenter, prospective, randomized trial at three children's hospitals in Shanghai in acute phase of KD. They were randomly divided into three groups: The group A was given IVIG 2 g/kg once via intravenous infusion, in 12 to 24 hours; the group B was given IVIG 1 g/kg twice via intravenous infusion over two days; and the group C was given IVIG 1 g/kg once via intravenous infusion. All the results were statistically analyzed.
Results: There were 138 cases in Group A, 127 cases in Group B, and 132 cases in Group C. During this groups, we found no difference in age and weight. Also there was no difference in the incidence of IKD, IVIG resistance and coronary arterial lesion. There was no difference in clinical signs and the laboratory datas before treatment. No difference in the febrile period before hospitalized in three groups. After treatment, there was no significant difference in three groups in the time of fever relieving. Also there was no significant difference in the duration of fever. there was no difference in the changes of these laboratory datas. Coronary arterial lesion occurred in 57 patients. No difference was observed in the incidence of coronary arterial lesion in three groups. No difference in the incidence of coronary arterial lesion during the follow up. The length of hospital stay and hospital costs satisfy a linear relationship, we found that the cost of Group C was the lowest while the Group B was the most expensive.
Conclusions: Administration of IVIG in a single dose of 1 g/kg in combination with aspirin can become a preferred treatment regimen for Kawasaki disease.

P1373 - SURGERY FOR RHEUMATIC HEART DISEASE A 5 YEAR (2010–2015) REVIEW FROM NAMIBIA

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Methods: Retrospective review of patients in the RHD Registry referred for cardiac surgery between October 2010 and December 2015. Case notes, echocardiograms and surgical reports were interrogated. Data captured using Microsoft Excel, coded and analyzed using STATA 14.0.
Results: Two hundred (200) of 536 patients had 205 valve operations. Females 125 (62.5%). One hundred seventy-one (83.4%) under 40 years and 26.3%, children. Sixteen had previous surgery. Patients had advanced disease; 81% NYHA Class III or IV, atrial fibrillation 74%, 70% pulmonary hypertension. Mitral valve surgery performed in 162; 35 repair and 127 replacement of which 60 bioprosthetic and 67 mechanical valves. Aortic position 72, 51 mechanical and 21 tissue valves. Sixty-six (66) patients double valve surgery (51 mitral-aortic, 15 mitral-tricuspid). Mean follow-up 33.98 months. Thirty-two patients (17%) died, 11 within 30 days. Amongst deaths, 12 only mitral valve, 3 only aortic valve, 11 aortic with mitral, 6 mitral and tricuspid surgery. Nineteen (59.4%) deaths occurred in patients younger than 30 years of age. Sixty month survival; mitral repair 90% vs mechanical 76% and aortic tissue 88% vs mechanical 76%. Of mitral valve repair 15 developed post-op regurgitation and 4 required re-operation. Aortic stenosis occurred in 19% tissue and 7.6% mechanical valves.
Conclusion: A large cohort needed surgery in a country previously unable to care for patients with RHD. Patients are young, very sick and predominantly female. High mortality reflects disease severity. Valve repair is preferred over replacement in RHD however, all aortic and many mitral valves required replacement. Difficulty with anticoagulation increases use of tissue rather than mechanical valves. Survival is improved with tissue valves but risk of stenosis is higher.

P1377 - RHEUMATIC HEART DISEASE IN CHILDREN UNDER FIVE YEARS AT THE UGANDA HEART INSTITUTE

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Introduction: Rheumatic heart disease (RHD) is the most serious complication of rheumatic fever that commonly affects children aged 5-15years. Children under five years are more likely to have severe carditis. Unfortunately primary prevention has been given less attention in this age group. We set out to describe case series of children under five years with RHD at the Uganda Heart Institute.
Methods: We conducted a retrospective chart review of children aged less than 5 years with a diagnosis of RHD seen at the Uganda Heart Institute from 2010 to 2016. All echocardiograms were reviewed by a paediatric cardiologist using the 2012 World Heart Federation criteria for the echocardiographic diagnosis of RHD.
Results: RHD was confirmed in 379 children less than 18 years. Seven children (1.8%) were under 5 years with a mean age of 2.5 years, range (2–4.5) years. All had advanced disease with severe mitral valve regurgitation and functional tricuspid valve regurgitation. Eighty five percent had moderate to severely elevated pulmonary pressures. None had congenital heart disease. One child had moderate mitral valve stenosis with no left sided obstructive lesions. We describe one case in detail; 2 year old male presented in congestive heart failure, with pedal oedema, ascites, displaced apex beat and grade3/6 holosystolic murmur at the apex.

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Echocardiogram revealed thickened mitral valve leaflets (3.8mm), severe mitral regurgitation with an eccentric jet, and mild tricuspid regurgitation PG 45 mmHg. Unfortunately, he died 2 years after diagnosis.
Conclusion: We share unique cases of RHD in children under 5 years who presented with severe disease at the Uganda Heart Institute. Screening programs should not be limited to school going children or those above 5 years. Genetic studies are recommended to assess the particular strains of group A streptococcus that cause severe disease in these children.

P1467 - IMPROVING DELIVERY OF SECONDARY PROPHYLAXIS TO PEOPLE LIVING WITH RHD IN FIJI ISLANDS

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Rheumatic Heart Disease (RHD) is a significant public health problem in Fiji Islands. Episodes of acute rheumatic fever (ARF) are often unrecognised and cases of RHD often present in severe stages. The Fiji Islands RHD Control and Prevention Project is delivering a four year Project led by Fiji Ministry of Health and Medical Services. The Project has implemented a series of interventions aimed at improving adherence by 1) improving knowledge of the disease among patients and carers 2) improving knowledge of health professionals regarding secondary prophylaxis treatment and importance of patient counselling 3) providing support to patients and carers. The Project piloted models of care at eleven sites with the planned wider roll out across the country. Stakeholder workshops and patient/carer interviews were conducted in the design and interim phases. The effectiveness of interventions were evaluated via the national rheumatic fever information system which records Benzathine penicillin adherence data. Qualitative and quantitative data were gathered and analysed for training, media campaign and patient and carer support activities. Support groups were conducted at health centres and led by nurses using educational flip charts followed by open discussion. The Project has reached over 600 patients and carers through support group activities. Approximately 45% of the nurse workforce were trained in 2016. Preliminary adherence data is showing an average of 20% improvement in adherence among ARF/RHD patients at pilot sites over a 12 month period and a multi- media campaign aimed at non-adherent patients was launched in 2016. The Project, has the potential to contribute to the development of a model of chronic disease care that can be applied in other high RHD prevalence countries.

P1518 - A RARE CASE OF LARGE PERICARDIAL EFFUSION WITH FIBRINOUS APPEARANCE IN ACUTE RHEUMATIC FEVER

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Background: Acute rheumatic fever is a non-suppurative complication that occurs following beta-hemolytic streptococci group A infection. Cardiac involvement may be in the form of pancarditis by involving each three layers of the heart. Pericardial involvement is mostly in the form of benign and mild pericadial effusion. Rarely, severe pericardial effusion and tamponade may occur.
Case Report: The authors describe a 13-year-old-girl who was diagnosed with rheumatic fever a year ago and was not using Penicilin for secondary prophylaxis. She was admitted to the hospital with fever and joint pain two days ago and shortness of breath 24 hours ago. On physical examination, she had a severe general condition, dyspneic, hypocorax, swollen ankle and hepatomegaly. Heart rate was 132bpm. In the thoracic auscultation, cardiac sounds were rhythmic and hypophonetic with a systolic murmur in the mitral region. Chest radiography revealed a considerable cardiomegaly and water bottle appearance. On the electrocardiogram, she presented sinus rhythm with signs of left ventricle enlargement and altered ventricular repolarization. The echocardiogram revealed severe mitral regurgitation and a large pericardial effusion with fibrinous strands traversing the pericardial space. She underwent venous corticotheraphy for ten days and then continued with oral doses. Oxacilin antibiotic was also started but was suspended nine days after. She evolved with excelente improvement of her symptoms and her echocardiogram nine days after revealed almost no pericardial effusion.
Conclusion: The authors draws attention to the uncommom presentation of the pericardial effusion in this case with large effusion and the aspect of the bundle of fibrinous strands in the pericardial space and for the good result with corticotheraphy, not requiring pericardial drainage.

P1574 - TRIPLE VALVE REPAIR IN YOUNG RHEUMATICS

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Rheumatic heart disease is a significant problem in developing countries and afflicts young children, in whom valve replacement is clearly not a good option. We present our experience in managing 5 young patients with aortic, mitral and tricuspid regurgitation who underwent repair of all the three valves. 5 young patients, all of them female between the age group 11 – 18 years were operated for aortic, mitral and tricuspid regurgitation. The aortic annulus was less than 16mm in all the patients(11–16), the mitral annulus had a mean size of 26 mm(23–31), the tricuspid regurgitation was moderate in all with moderate pulmonary artery hypertension. The method used was leaflet placation, bicuspidisation, leaflet extension and thinning of leaflets in pts with aortic regurgitation. Mitral valve was repaired by anterior leaflet chordal shortening by artificial chords or cusp level chordal shortening and the PML was extended by autologous pericardium and a partial posterior annuloplasty. Tricuspid valve was repaired by sectoral annuloplasty along the anterior posterior area of the annulus. The mean cross clamp time was 110 minutes and bypass time was 140 minutes. Del Nido cardioplegia with moderate hypothermia was used in all. 3 patients no AR, two had mild + AR, 4 patients had no MR. The Post op course was uncomplicated in all the patients and all the patients could be shifted out of ICU by 3rd day and discharged by 6 days. Triple valve repair provides satisfactory outcome atleast in the short term. Leaflet extension of all three leaflets or aortic valve with PTFE membrane mitral valve pathology with anterior leaflet prolapse and restricted posterior leaflet motion have the best result.

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P1601 - THROMBOSPONDIN 2 PREDICT WHETHER CHILDREN WITH KAWASAKI DISEASE RESPOND TO TREATMENT WITH INTRAVENOUS IMMUNOGLOBULIN

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Object: The present study was to investigate the predictive value of thrombospondin-2(TSP-2) in assessing the response to intravenous immunoglobulin (IVIG) in children with acute Kawasaki disease (KD).
Methods: 61 children with KD were recruited as KD group, and healthy children (n = 32) and febrile children (n = 32) were used as control groups. ELISA was used to measure plasma TSP-2 and TSP-1 levels.
Results: Compared with the control groups, the plasma TSP-2 and TSP-1 levels in acute KD were significantly elevated (TSP-2: 30.56 ± 11.32 vs 21.35 ± 8.82 vs 16.37 ± 2.86ng/ml, P > 0.001; TSP-1: 10271.33 ± 7538.38 vs 7174.31 ± 4807.37 vs 3660.51 ± 2671.90ng/ml, P < 0.001). The plasma TSP-2 level in the IVIG non-responders (13/61) was significantly higher than the responder group (48/61) (38.59 ± 12.12 vs 28.39 ± 10.17ng/ml, P = 0.003). When using an receiver operating characteristic (ROC) curve to analyze the predictive effect of TSP-2 on non-responsiveness to IVIG treatment, the area under the curve(AUC) was 0.776(95% CI: 0.640 to 0.913; P = 0.002). When the cutoff value for TSP-2 was 31.76 ng/ml, the sensitivity was 76.90%, the specificity was 70.80%.
Conclusion: The plasma TSP-2 level was elevated in acute KD and predicted the therapeutic response of children with KD to IVIG with high sensitivity and specificity, which could help guide clinical decisions and lead to better early treatment to prevent complications.

P1633 - EFFICACY OF FOUR SCORING SYSTEMS IN PREDICTING INTRAVENOUS IMMUNOGLOBULIN RESISTANCE IN CHILDREN WITH KAWASAKI DISEASE IN A CHILDREN'S HOSPITAL IN BEIJING NORTH CHINA

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Objective: To evaluate the predictive efficacies of four existing scoring systems for intravenous immunoglobulin (IVIG)-resistance in Kawasaki disease (KD) in hospitalized children with KD in Children's Hospital affiliated to Capital Institute of Pediatrics, Beijing, China.
Study Design: We retrospectively analyzed 1569 cases with KD treated at our children's hospital from January 2010 to December 2015. Age, gender, clinical manifestations, and pretreatment hematologic indicators were recorded. Scores were assigned using four existing scoring systems: Egami, Kobayashi, San Diego, and Formosa system. A four case table test was used to determine prediction efficacies.
Results: There were 63 IVIG-resistant cases (41 males, 22 females; average age, 2.5 years). Nine cases were classified as high risk for IVIG resistance by Egami, and this system had a sensitivity and specificity of 14% and 86%, respectively. Ten cases had Kobayashi high-risk scores, and this system had a sensitivity and specificity of 16% and 85%, respectively. The San Diego system assigned 60 cases as high-risk, and the sensitivity and specificity of this system was 95% and 3%, respectively. Finally, 27 cases had Formosa scores in the high-risk category, and the sensitivity and specificity were 43% and 47%, respectively.
Conclusions: None of the evaluated systems to assess risk for IVIG resistance displayed the combination of sensitivity and specificity necessary for screening. Our analyses show that the four scoring systems have limited utility in predicting IVIG-resistance among patients with KD in our population.

P1657 - CHARACTERISTICS OF RHEUMATIC HEART DISEASE IN MANITOBA CANADA A 10 YEAR RETROSPECTIVE STUDY

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Objective: As previously reported, Acute Rheumatic Fever (ARF) remains a significant health problem between the First Nation children of Manitoba. The aims and objectives of this study are to determine the characteristics of Rheumatic Heart Disease (RHD) present in children affected by the disease in the Province of Manitoba, Canada.
Methods: Retrospective study conducted at the Variety Heart Centre, Winnipeg, MB, Canada. Children with discharged diagnosis of ARF were identified through the Children's Hospital of Winnipeg medical records and the Electronic Echocardiography data based at the Variety Heart Centre.
Results: Over a 10-year study period (2000-2010), a total of 42 [male:23, female: 19, median age 9.6 years (min 3.9 max14.9)] patients meeting the Jones criteria for ARF and evidence of RHD are included in this study. Echocardiographic follow up period more than 12 months was the major criterion for enrolment in the study: [median 4.99 years (max 11 and min 1.45 years)]. Marked RHD at presentation defined as moderate or severe mitral (13 and 11 cases) or aortic (2 and 1 cases) insufficiency. Moderate or severe insufficiency affecting both of the valves was observed in 3 patients. Moderate or severe mitral valve insufficiency at follow up was present in 6 and 4 patients respectively, with similar aortic valve findings in 2 patients from each group. Four (4) patients required either mitral or aortic prosthetic valve. Decreased function at presentation was detected in 2 patients and remained decreased in one of them even after valve replacement. There were no deaths related to RHD.
Conclusion: RHD remains a significant health problem between the First Nation children of Manitoba. Therefore, a strategy of primary prevention of ARF based on health education, improvement of social conditions, access to care, early diagnosis and effective treatment of the disease should be implemented.

P1771 - A CASE OF INFANTILE RUPTURED MITRAL CHORDAE TENDINEAE COMPLICATING INCOMPLETE KAWASAKI DISEASE

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Background: Infantile ruptured mitral chordae tendineae is a rare disease causing acute cardiopulmonary failure. We encountered a patient with concomitant ruptured mitral chordae tendineae with incomplete Kawasaki disease with no main symptoms other than fever, which declined spontaneously.
Case Presentation: The patient was a 7-month-old girl who developed fever (38-39 °C) of unknown origin. The fever persisted for 6 days and then declined spontaneously (36-37 °C). There were no other main symptoms of Kawasaki disease and no flare was noted in the BCG region. The amount ingested and the urinary volume decreased from about 12 days after the development of fever, and the patient was brought to our hospital on day 14. Gallop rhythm and heart murmur were heard. Echocardiography was performed, and severe mitral regurgitation due to ruptured chordae tendineae and coronary arterial dilation (Z-score: RCA, 3.51; LMCA, 5.40; LAD, 5.31) were observed. In laboratory data, increases in C-reactive protein and brain natriuretic peptide, anemia, thrombocytosis, and hypoalbuminemia were observed. Based on the coronary arterial lesion and laboratory data, the patient was diagnosed with valvular impairment complicating incomplete Kawasaki disease. The inflammatory reaction persisted and was treated with intravenous immunoglobulin and steroids, followed by mitral annuloplasty. The postoperative course was favorable and the coronary arterial lesion improved.
Conclusion: Serious valvular impairment, such as ruptured mitral chordae tendineae, occurs even in cases of incomplete Kawasaki disease, i.e., in the absence of the main symptoms of Kawasaki disease, and in which fever declines spontaneously.

P1784 - EPIDEMIOLOGY OF ACUTE RHEUMATIC FEVER AND KAWASAKI DISEASE IN HAWAI'I BETWEEN 2009 AND 2015

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Background: Kawasaki disease (KD) is currently estimated to be the most common cause of acquired heart disease in children in the developed world, while rheumatic fever (RF) is the most common cause of acquired heart disease in the developing world. Historically, Hawai'i has had a higher incidence of both conditions compared to the continental United States but recent Hawaii specific demographic data are lacking.
Methods: A data base review using discharge ICD 9 diagnosis codes was conducted to identify all patients between 0-18 years of age who were hospitalized for acute RF or KD at two nonmilitary children's hospitals between 2009 and 2015. Only the initial hospitalization was counted. Basic information included age at diagnosis, gender and ethnicity. Hawaii census data were used to calculate incidences.
Results: There were 336 total KD hospitalizations (mean age 2.6 years, 49% male, 82% <5 years), the overall average annual incidence was 17/100000 <18 years of age and 47/100000 <5 years of age. Incidence was highest among Asian children <5 years (52/100000), whereas Caucasian children <5 only had an incidence of 9/100000. There was no noticeable seasonality. For RF, the average incidence was 5/100000 <18 years of age (mean age 11.5 years, 60% male). Of these, 80% were of Hawaiian/Pacific Islander ethnicity, giving an incidence of 41/100000 in this subpopulation and 67% of patients were between 10-16 years at diagnosis.
Conclusion: Between 2009 and 2015, the overall incidence of KD and RF was 2 to 2.5 times higher than described historically in the continental US but similar compared to historic cohorts in Hawaii. Asian children under 5 years are at particular risk for KD and older children of Hawaiian/Pacific Islander ethnicity predominated in the RF cohort, confirming epidemiologic data from previous decades.

P1859 - KAWASAKI DISEASE ANALISIS OF FACTORS ASSOCIATED WITH CARDIOVASCULAR LESIONS

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Sant Joan De Déu Barcelona Children´s Hospital, Pediatric Cardiology, Barcelona-Spain[1]; Miguel Servet Hospital, Pediatric Cardiology, Zaragoza-Spain[2]

Introduction: Kawasaki disease (KD) is the most common cause of acquired heart disease in childhood. Clinical and laboratory features along with treatment strategies have been associated with development of cardiovascular lesions. This study aims to determine the incidence of cardiovascular lesions and to analyze clinical, laboratory and treatment factors associated with the development of cardiovascular lesions.
Material and Methods: Patients diagnosed with KD in two University Tertiary Hospitals (Barcelona and Zaragoza) between 2013 and 2015 were retrospectively analyzed. The Japanese Society of Pediatric Cardiology clinical guidelines were used for diagnosis and classification. Demographics, clinical and laboratory variables and the presence of cardiovascular abnormalities were collected. Data are presented as median (IQR). SPSS® was used for the analysis of statistical data.
Results: Fifty-eight children were included, 35 (60.3%) males, aged 24 months (12-48). Thirty-two (55.2%) were complete; 19 (32.8%) incomplete and 7 (12.1%) atypical forms of KD. Cardiovascular abnormalities were found in 18 (30.5%): mitral regurgitation 11 (19%), myocardial dysfunction 3 (5.2%), pericardial effusion 3 (5.2%) and coronary aneurysms 8 (13.6%). Patients with more than 7 days of fever had more coronary aneurysms (75% vs 38% $p = 0.06$), although it was not statistically significant. Incomplete and atypical presentations associated more cardiovascular abnormalities compared with complete KD (37.5% - 62.5% vs. 0%, $p < 0.01$) specifically coronary aneurysms (38.9% - 38.9% vs 22.2%, $p < 0.05$).There was no relationship with laboratory features. Children with delayed IVIG treatment (10 days after KD onset) had more coronary aneurysms (50% vs 12% $p = 0.02$).
Conclusion: Duration of fever, type of presentation of KD and the time of administration of IVIG are proposed as markers of cardiovascular risk and coronary aneurysms.

P1896 - DEMOGRAPHIC CLINICAL AND ECHOCARDIOGRAPHIC FEATURES OF 871 PATIENTS WITH RHEUMATIC HEART DISEASE THE RHEUMATIC HEART DISEASE REGISTER

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Background: Rheumatic Heart Disease (RHD) is an important public health problem in Sudan with prevalence of 10:1000. A Register was established in order to monitor RHD-related health outcomes towards achievement of World Heart Federation objective of 25% reduction in mortality of under 25yrs by 2025. Data collected from register will periodically be analyzed to better understand impact of RHD on various states of Sudan and thus enable effective planning of services and management of resources.
Materials & Methods: A Register was initiated in two hospitals (Jaffar Ibn Ouf Children's Specialized Hospital and Sudan Heart Center) since January 2006. All patients with clinical/echocardiographic evidence of RHD were included. Their demographic data, clinical/echo findings, and contact details were recorded. Data covered period January-2006 to October-2016.
Results: 871 patients were reviewed (54% Male). Age ranged 4 to 25 years. Highest incidence (55%) was observed in 10 to 15 years old. 26% of registered population was from Darfur, 25% from Kurdufan, 12% from Al Jazirah, while Northern and Eastern areas showed less affection (Figure-1). Subsequent comparison of cases/100,000 on basis of state's population (Table-1) revealed that White Nile had highest RHD rate per capita (3.1/100,000), followed by Kurdufan (2.8/100,000), then Sinnar (1.8/100,000),

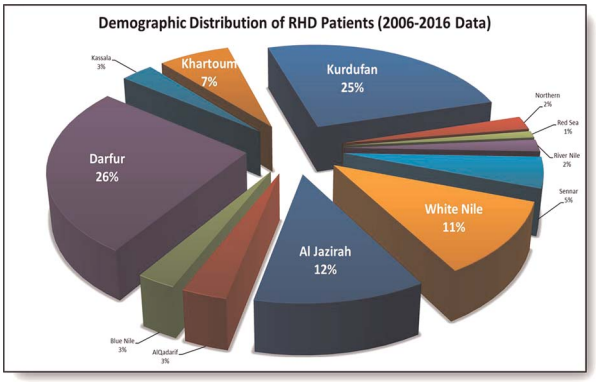

Figure 1.

Table 1. Demographic Distribution of Patients with Severe form of RHD

State	No. of Cases	Percentage
Kurdufan	76	29.3%
Darfur	67	25.9%
White Nile	21	8.1%
Khartoum	19	7.3%
Al Jazirah	16	6.2%
Al Qadarif	9	3.5%
Kassala	9	3.5%
Blue Nile	8	3.1%
Northern	6	2.3%
Sennar	5	1.9%
River Nile	3	1.2%
Red Sea	2	0.8%
Others	18	6.9%
	259	100%

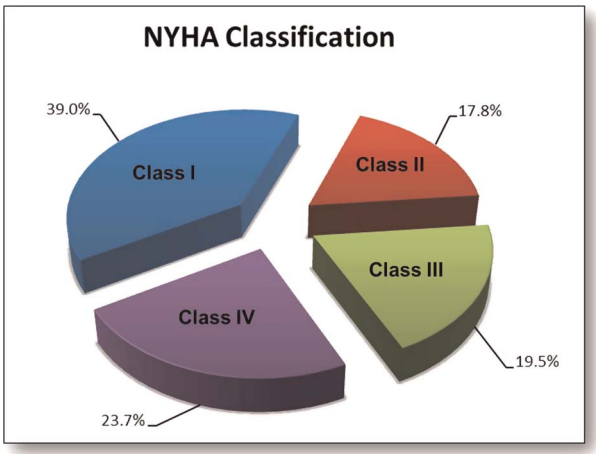

Figure 2.

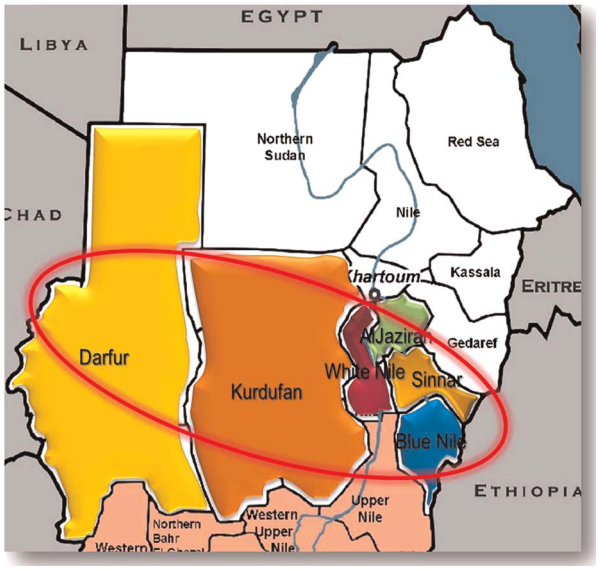

Figure 3.

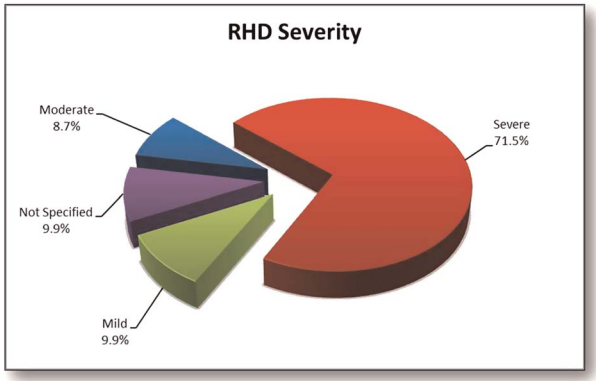

Figure 4.

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then Darfur (1.7/100,000). The study revealed an apparent RHD belt extending from Darfur to Blue Nile (Figure-2). Echo revealed mitral regurgitation (MR) in 78.1%, isolated aortic regurgitation (AR) in 2.7%, and >2 lesions in 15.4% (Table-2 and Figures 3 & 4). In terms of demographic distribution of 259 patients with severe RHD, 29.3% were from Kurdufan, 25.9% from Darfur, and 8.1% from White Nile (Table-3).
Conclusion: There is an apparent RHD belt extending from Darfur to Blue Nile. RHD patients having severe valve affection were mostly from Kurdufan and Darfur. Presentation is late with the severe disease form as it is not caught at acute phase.

Table 2. Residency Distribution Vs RHD Cases per 100,000

State Residency	Population	No. of Cases	Cases per 100.000
White Nile	1,730,588	54	3.12
Kurdufan	4,327,396	120	2.77
Sennar	1,285,058	23	1.79
Darfur	7,515,445	125	1.66
Al Jazlrah	3,575,280	56	1.57
Blue Nile	832,112	13	1.56
Other States	11,628,121	5 to 15	<1.5
Overall	30,894,000	478	1.55

Table 3. Valve Lesions Percentages of RHD patients

Valve Lesions	No. of Cases	Percentage
MR	256	31.9%
AR	22	2.7%
MS	16	2.0%
TR	0	0.0%
MR/AR	293	36.5%
MR/ MS	37	4.6%
MR/TR	41	5.1%
AR/MS	6	0.7%
AR/TR	6	0.7%
MS/TR	2	0.2%
More than 2 lesions	124	15.4%
	803	100%

P1913 - KAWASAKI DISEASE ITS MANIFESTATION AFTER SIMULTANEOUS IMMUNIZATION WITH MEASLES RUBELLA VARICELLA AND PNEUMOCOCCAL VACCINE SUGGESTING ITS POSSIBLE ASSOCIATION WITH MEASLES VACCINATION

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Background: Although some previous reports suggested the possible association between vaccination and Kawasaki disease (KD), a recent population-based study showed vaccines did not increase, but rather decreased the incidence of KD shortly after vaccination. We report a pediatric patient: KD manifested just after simultaneous immunization with measles-rubella, varicella, and pneumococcus vaccine. Vaccination, especially measles vaccination, was suggested to be associated with KD ocurrence.
Case: A 14-month-old Japanese girl had a fever the day after concomitant inoculation with measles–rubella (MR) (first dose), varicella (first), and pneumococcal (fourth) vaccination. On day 2, rash appeared around the previous BCG inoculation site. On day 4, her condition met 4/6 KD diagnostic criteria (CRP 4.24 mg/dL, WBC 18200 /μL). On day 5, on meeting 5/6 KD criteria, she was diagnosed with complete KD. KD manifestations resolved with decreases in CRP/ WBC. However, rash around the BCG inoculation site was still evident, which became a crust on Day 11. Aspirin was solely administered. Antibody analysis on Day 6 showed: rubella(-), VZV(-), and EBV(-), and importantly, measles IgG(-)/IgM(+). Ferver did not recur. Ultrasound revealed no coronary artery sequelae throughout her course.
Discussion and Conclusion: KD appeared just after concomitant immunization with MR, varicella, and pneumococcal vaccine. Rash around the previous BCG site is pathognomonic for KD; this strongly suggests this patient had KD. Fever due to MR vaccination usually occurs on day 3 to 12 after inoculation, when viral multiplication occurs within the host. In this patient, fever occurred much earlier (day 1), reducing the possibility of fever due to live-vaccination. The measles IgG(-)/IgM(+) obtained on Day 6 suggested that the measles virus inoculation may have caused, or at least been associated with, KD. Further monitoring may be needed to clarify the association between measles vaccination and KD.

P1947 - VASCULITIS RESULTING IN GIANT CORONARY ARTERY ANEURYSMS AND SYSTEMIC MULTIPLE ANEURYSMS IN A FIVE MONTH OLD INFANT

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Background: Systemic vasculitis is a group of diseases characterized by the inflammation of blood vessels. Among the primary vasculitis, Kawasaki disease is common and occurs almost exclusively in childhood. It predominantly affects medium-sized arteries, most commonly the coronary arteries. Vasculitis predominantly affecting large vessels in childhood is Takayasu arteritis, which mostly involves the aorta and its major branches and can also affects some medium-sized arteries.
Materials and Methods: A five-month-old infant had recurrent fever for more than one month. The echocardiography indicated giant coronary artery aneurysm with continuous progress. The MRI imaging presented systemic multiple aneurysms, including aneurysms of subclavian, axillary, brachial, abdominal aorta, iliac and Superior mesenteric arteries, and dilated carotid, radial, femoral and celiac trunk arteries. The child was successively treated with high-dose intravenous immuneglobulin, glucocorticosteroid pulse, cyclophosphamide pulse and TNF-α antagonist (infliximab). However finally he died.
Results: An autopsy was performed with the parental consents. The autopsy indicated that the child died because of the rupture of coronary artery aneurysm and pericardial tamponade. Different types of blood vessels were sampled and histopathological examination confirmed vasculitis of large- and medium-sized arteries, and normal small-sized arteries.
Conclusions: The child was finally diagnosed with large- and medium-size vessel vasculitis, inclining to Kawasaki disease. Although we have tried multiple medications, the severity of coronary arteries aneurysms finally determined the prognosis of the disease.

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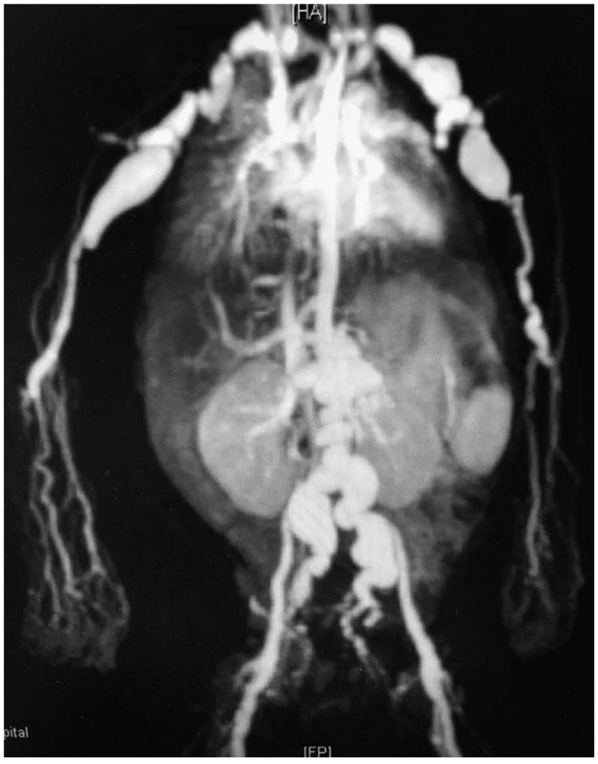

Figure.

P1997 - THE USEFULNESS AND SERIAL CHANGES IN NEUTROPHIL TO LYMPHOCYTE RATIO AND PLATELET TO LYMPHOCYTE RATIO IN KAWASAKI DISEASE

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Background: Kawasaki disease is an inflammatory Condition which needs to be distinguished from other infectious diseases. The white blood cell count and its subpopulations in the peripheral blood are classic indicators of inflammation. Neutrophil-to-lymphocyte ratio (NLR) and platelet-to-lymphocyte ratio (PLR) are useful as systemic inflammatory markers and prognostic indicators of adverse cardiovascular events and cancers. Also, they may provide additional information to conventional markers for predicting IVIG resistance in patients with KD. The aim of the study is to evaluate usefulness of NLR and PLR in diagnosis of Kawasaki among other infectious disease causing fever. Also, serial changes of NLR and PLR in Kawasaki disease throughout the disease course.
Methods: Ninety six children with Kawasaki disease and 41 controls were retrospectively enrolled. Their leukocyte, neutrophil, lymphocyte counts and platelet counts were recorded serially, before IVIG (D0), 2 days after IVIG (D2), 14 days (D14) and 56 days (D56) after IVIG treatment.
Results: Leukocyte [$p = 0.002$], neutrophil [$p = 0.001$], and platelet [$p < 0.001$] counts were significantly higher in Kawasaki disease patients compared to controls. NLR [$p = 0.02$] was also significantly higher in Kawasaki disease, however, there was no significant difference regarding PLR [$p = 0.291$]. On serial measurement of NLR and PLR in patients with Kawasaki disease, NLR dropped significantly and continuously after IVIG treatment. (D0 vs D2, D0 vs D14, D0 vs D56, all p values are <0.001). On the other hand, PLR significantly increased after IVIG treatment (D0 vs D2, $p < 0.001$), then dropped afterwards (D0 vs D14, D0 vs D56, all p values <0.001)
Conclusion: Neutrophil to lymphocyte ratio values was useful in diagnosing Kawasaki disease from other infectious diseases however, unlike other studies, platelet to lymphocyte ratio was not as useful. In addition, NLR significantly and continuously drops after IVIG treatment.

P2077 - DOPPLER ECHOCARDIOGRAPHY FOR SUBCLINICAL RHEUMATIC HEART DISEASE EVALUATION OF A COMPUTERISED DIAGNOSIS OF THE MITRAL VALVE APPARATUS

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Background: Rheumatic Fever and Rheumatic Heart Disease remain a major burden among children in developing countries. Echocardiography with colour flow Doppler is key to early diagnosis, especially in subclinical cases. Jones criteria revision recently included echocardiography screening as an important criterion for diagnosis. However, the technique requires time and experienced operators, which are scarce resources in the affected areas. A computer-aided analysis of the echocardiography images could, potentially, reduce the cost of screening, and spread diagnostic accessibility for a larger number of patients. Ongoing developments on the semi-automatic mitral regurgitation jet detection are presented. These results are part of a major project (NORTE-01-0247-FEDER--003507-RHDecho, co-funded by PORTUGAL2020) which also aims to track the mitral valve leaflets on brightness mode echocardiography.
Materials and Methods: The implemented algorithm starts by finding a region containing the left atrium, given an input from the user (centre of the chamber). Then, the colour patterns which are congruent with reflux are automatically selected as mitral regurgitation candidates. A paediatric screening campaign was conducted by the Non-governmental organization *Círculo do Coração* at the state of Paraíba, Brazil. A total of 979 frames (apical 4 chamber view and parasternal long axis view) from 20 patients showing echocardiographic findings of mitral valve insufficiency were manually annotated and used to validate the proposed methods.
Results: Overall, results for colour pattern detection are promising (sensitivity = 0.92 false detection rate = 0.10), but further developments are required for left atrium delineation (sensitivity = 0.82, false detection rate = 0.25).
Conclusions: The proposed computational algorithms present promising results to be part of an impactful tool for the early diagnosis of subclinical rheumatic heart disease. The collaboration between doctors and engineers can, potentially, revolutionize the way diagnosis of such serious diseases is made.

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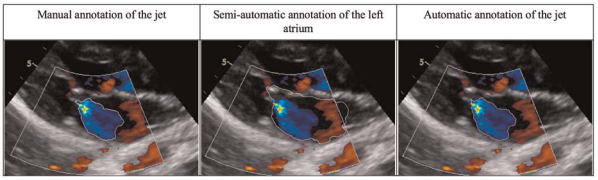

Figure.

P2095 - PHYSICIANS HEALTH CARE WORKERS AND THE PUBLIC HOW TO TEACH ABOUT ARF AND RHD

Alyaa Kotby
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Control of Acute Rheumatic fever (ARF) and Rheumatic heart disease (RHD) entails the development and implementation of a comprehensive strategy for teaching physicians, health workers and the public. Undergraduate teaching of ARF and RHD to medical students and nursing staff should be implemented not only in moderate and high risk communities but also in low risk countries since resurgence of RF can occur due to changing epidemiological and virulence factors. A two level approach should be adopted in physician training. The first should give the basic knowledge of how to identify and treat streptococcal throat infection, suspect and investigate RF and RHD. The target physicians include all general practitioners, family doctors, pediatricians and cardiologists. Second level training is for physicians in specialized referral centers for management of notified cases of ARF and RHD. Physicians and health care workers should be guided by the local community guidelines tailored for management in their community. Complete collaboration between the ministries of Health and Education and advocacy insures a continuous awareness campaign for the public. Campaigns include all stakeholders with government representatives, politicians, teachers, religion figures, pharmacists, football players, singers and actors. Films, cartoons and stories with local heroes are used to increase the awareness and compliance. Multimodal approaches to convey the information in a culturally appropriate manner should use verbal and popular media available in each community. Continuous information is sent in simple terms till the whole community is talking about ARF and RHD. Universal teaching will ultimately lead to universal end of ARF and RHD.

P2103 - STRENGTHENING CARDIOVASCULAR CARE WITHOUT SPECIALISTS INTRODUCTION OF HANDHELD ECHOCARDIOGRAPHY TO FACILITATE IMPROVED DIAGNOSIS OF PEDIATRIC HEART FAILURE IN A LOW RESOURCE SETTING

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Background: Heart failure (HF) is an important cause of morbidity and mortality in Ugandan children. Access to cardiovascular care is limited. The objective this study was to determine the pattern of pediatric heart failure diagnosis prior to and after introduction of handheld echocardiography.
Materials and Methods: Inclusion criteria included age $\geq$ 5 years and at least 1 of 10 pre-determined signs of HF. An attempt was made to capture all eligible patients in the outpatient, inpatient, and emergency departments. In phase 1 (4-months) final diagnosis was made according to clinical evaluation. In phase 2 (currently in month-2) handheld echocardiography used by local healthcare providers was introduced to facilitate improved diagnostic accuracy.
Results: To date (phase 2 ongoing), 847 patients have been enrolled. Of these, 86 (10%) are children (11.7 ± 4.0 years; 57% female). The most frequent pediatric enrollment criteria include dyspnea (80), high provider suspicion of CV disease (70), and tachycardia (42). Phase 1 enrolled 65 children with 54 (83%) having a final diagnosis of HF (2-pericardial effusion, 1-hypertensive HF, 6-congenital, 27-rheumatic, 2-other valvular, 16 HF NOS). Phase 2 has enrolled 21 patients with 9 (43%) having a final diagnosis of HF (1 pericardial effusion, 3 rheumatic, 4 dilated cardiomyopathy, 1 right-sided HF). Echocardiography appears to improve diagnostic specificity for HF (86% vs. 43% of suspected children), though not yet statistically significant ($p = 0.15$).
Conclusions: Approximately 10% of patients presenting at a Ugandan regional hospital with possible HF are children. Preliminary data suggests that echocardiography improves the specificity of this diagnosis. Further data collection and analysis is needed to fully understand the epidemiology of pediatric HF and to determine the impact of handheld echocardiography to improve diagnostic accuracy and outcomes in Uganda and other low-resource settings.

P2104 - LONGITUDINAL OUTCOMES OF UGANDAN CHILDREN IDENTIFIED WITH LATENT RHEUMATIC HEART DISEASE THROUGH SCHOOL BASED SCREENING

Andrea Beaton[1], *Emmy Okello*[2], *Twalib Aliku*[3], *Joselyn Rwebembera*[2], *Grace Mirembe*[4], *Amy Scheel*[1], *Alyssa Dewyer*[1], *Chris Longenecker*[5], *Craig Sable*[1], *Peter Lwabi*[2]
Children's National Medical Center, Cardiology, Washington DC-United States[1]; *The Uganda Heart Institute, Cardiology, Kampala-Uganda*[2]; *Gulu Regional Referral Center, Cardiology, Gulu-Uganda*[3]; *The Joint Clinical Research Center, Medicine, Lubowa-Uganda*[4]; *Case Western Reserve University, Cardiology, Cleveland OH-United States*[5]

Background: The importance of echocardiographic screening for latent rheumatic heart disease (RHD) is not known. Longitudinal data on these populations are limited. We report a large series of children with latent RHD and establish a consistent reporting framework to improve interpretation and generalization of data.
Materials and Methods: Children diagnosed with latent RHD (2010-2015) were enrolled in the Ugandan RHD Registry. Baseline echocardiograms were blindly re-reported (2012 WHF criteria) with subsequent echocardiograms reported in series. Definite RHD was sub-classified by severity. Progression and regression were precisely defined. Penicillin prophylaxis was recorded as a continuous variable when possible.
Results: Echocardiographic inclusion criteria was met by 227 out of 303 children (mean follow-up 2.9 years, SD 2.7 years): 164 borderline RHD (9 borderline-A, 135 borderline-B, 20 borderline-C), 43 with mild definite RHD (33 definite-A, 3 definite-C, 7 definite-D) & 20 with > mild definite RHD (14 definite-A, 3 definite-B, 3 definite-C). Seventy-six (46%) children with

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borderline RHD regressed, 72 (44%) were stable, and 16 (10%) progressed. Eighteen (42%) children with mild definite RHD regressed, 13 (30%) were stable, and 12 (28%) progressed. Outcomes were notably worse for children with >mild definite RHD, with only 2(10%) showing regression, 11 (55%) stable with moderate-to-severe RHD, and 7 (35%) with progression, including 3 deaths.
Conclusions: There is a large degree of heterogeneity in outcomes among children with latent RHD. Further analysis of this data is needed to determine the impact of penicillin prophylaxis, socioeconomic and demographic variables, and disease categorization on the trajectory of latent RHD. It is clear that children with greater than mild definite RHD should be considered missed clinical cases, as outcomes for these children are poor, and similar to published data on clinical RHD cohorts.

P2198 - A PROTOCOL FOR A SYSTEMATIC REVIEW OF THE DIAGNOSTIC ACCURACY OF HAND HELD ECHOCARDIOGRAPHY FOR THE DETECTION OF RHEUMATIC HEART DISEASE IN SCHOOL AGED CHILDREN AND ADOLESCENTS

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Background: Hand-held echocardiography presents an opportunity to address the need for cost-effective methods of diagnosing rheumatic heart disease (RHD) in developing countries where the disease carries high morbidity and mortality. Studies have demonstrated moderate sensitivity as well as high specificity and diagnostic odds for detecting latent RHD. We describe a protocol for systematic review of published primary diagnostic test accuracy studies to evaluate the evidence for this portable technology in diagnosing suspected RHD.
Methods/Design: Electronic search strategies will be conducted among various data sources including PubMed, Scopus, ISI Web of Science and EbscoHost. Primary observational studies of diagnostic accuracy of hand-held echocardiography versus standard echocardiography will be selected. Data extraction will be undertaken by two reviewers independently, to assess the methodological validity and quality of each study against QUADAS-2 criteria. Also, information relating to metrics of diagnostic accuracy and demographics will be extracted. Forest plots of sensitivity and specificity as well as a scatter plot in Receiver Operating Characteristic (ROC) space will be used to investigate heterogeneity of studies. If possible, a meta-analysis will be conducted to produce summary results of sensitivity and specificity using the Hierarchical Summary Receiver Operating Characteristic (HSROC) method. Subgroup analyses will include age, gender, geographical location and expertise of echocardiographic interpreter. Finally, a sensitivity analysis to investigate the effect of studies with a high risk of bias will be undertaken.
Discussion: This will be the first scientifically rigorous and comprehensive systematic review in the field, and may feed into evidence-based guidelines. Upon completion of this review, a paper will be submitted to a leading journal in the relevant field. Likewise, should the findings of this review warrant a change in clinical practice, a one page summary report will be disseminated among leading clinicians and healthcare professionals in the field. [Prospero Registration: CRD42016051261]

P2208 - ECHOCARDIOGRAPHIC FINDINGS OF CHILDREN DIAGNOSED WITH KAWASAKI DISEASE AND THEM EPIDEMIOLOGICAL AND CLINICAL FEATURES IN A REFERENCE HOSPITAL IN BOGOTÁ 2009 2014

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Background: Describe the echocardiographic findings, epidemiological and clinical features of patients diagnosed with Kawasaki disease (KD) in the period between 2009 and 2014 in a reference institution in Bogotá, Colombia.
Materials and Methods: This is an observational, cross-sectional study. Statistical analysis was made using Microsoft Excel 2010 and SPSS-22. The study population was obtained from the database of echocardiograms, reviewed the medical records and tabulated the information.
Results: Patients was 58, 6 patients didn´t accomplish inclusion criteria, total universe 52. 51.9% (27/52) female. The mean age of presentation was 33.4 months (interquartile range IR 36). Clinical features for the diagnosis of KD: 86.5% (45/52) of the patients had fever more than 5 days, 67.3% (35/52) had complete presentation. Clinical laboratory criteria: PCR elevation over than 30 mg/dl was found in 80% of the patients, high aminotransferases in 55%, leukocytes >15.000: 48%, normochromic normocytic anemia 36.5%, thrombocytosis 42.3%, albumin <3 g/dl 52%. The echocardiographic findings were: coronary artery aneurysms 9 patients (17.3%), coronary ectasia 2 cases (3.8%), and 78% of the patients had no pathological changes in the coronary arteries. Treatment: 92.3% (48/52) received acetylsalicylic acid, 100% received immunoglobulin. Regarding the risk factors for the development of coronary aneurysms, it was evidenced that in this cohort of patients studied the sex, complete presentation, hematocrit > 35% and leukocytosis > 12000 had a significant correlation.
Conclusions: KD is one of the most frequent vasculitis in childhood, predominantly involves the coronary arteries, but may also affect other large and medium-sized blood vessels; the etiology and pathophysiology have not yet been completely elucidated. However, their presentation suggests an interaction between genetic predisposition and environmental factors that may have infectious origin. It is a challenge for the pediatrician because the non-specific symptoms, but especially because early diagnosis and early initiation of treatment prevents the development of potentially fatal complications.

P2216 - FIFTY CASES OF CONTINUOUS INTRAVENOUS INFUSION OF CYCLOSPORINE A FOR PATIENTS WITH INTRAVENOUS IMMUNOGLOBULIN RESISTANT KAWASAKI DISEASE

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Background: Cyclosporin A (CsA) has been used to treat patients with intravenous immunoglobulin (IVIG)-resistant Kawasaki

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disease (KD); however, the major administration route of CsA reported in the literature has not been IV but oral. We reported 50 cases demonstrating the effects of continuous IV infusion of CsA (CICsA).
Materials and Methods: Fifty patients with KD (33 boys and 17 girls; age range, 3–110 months) who did not respond to repeated IVIG (≥4 g/kg) were enrolled. All the patients were started on CICsA (3 mg/kg/day; optimal serum level, 300–500 ng/mL) and then subsequently switched to oral CsA (4–6 mg/kg/day) approximately 1 week after their fever had subsided.
Results: Thirty-six of the 50 patients (72%) became afebrile within 24 h after CICsA treatment (range, ≤24 h to 7 days; median, 24 h). The CsA concentration at the time of defervescence was 220–570 ng/mL (median 373 ng/mL). The patients who were treated with CICsA before day 9 had no coronary artery aneurysm (CAA); however, one case of CAA (medium sides) was observed in a patient who had already experienced CAA on admission at day 10. There was no adverse effect of hypertension or hyperkalemia. The duration of CICsA was 4–27 days (median 8 days), and the total duration including internal use was 6–138 days (median 21 days). The serum C-reactive protein (CRP) and soluble interleukin-2R (sIL-2R) levels were significantly lower 3–5 days after CICsA (CRP: 13.1 to 4.4 mg/dL; sIL-2R: 2587 to 1638 U/mL, $p < 0.01$). The white blood cell count was not significantly changed from 16750 to 14048/μL, whereas the lymphocyte count was significantly changed (3021 to 4720/μL, $p < 0.01$).
Conclusions: CICsA was effective against IVIG-resistant KD, especially when the treatment was initiated before day 9, without serious adverse effects. Therefore, CICsA might be a good therapeutic option for treating IVIG-resistant KD.

P2241 - THE BURDEN OF RHEUMATIC HEART DISEASE IN AN ETHIOPIAN POPULATION FROM THE GONDAR REGION

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Background/Hypothesis: The prevalence rate of rheumatic heart disease (RHD) in Africa is 5-10/1000 school children. Little is known for the Gondar region, Ethiopia. The mass immigration of the Jewish population from the Gondar region to Israel created an opportunity for further investigation.
Materials/Methods: This cross-sectional study utilized a database of all recruits from a North-East Israel National Service Recruiting Center which included the medical history, physical examination findings together with cardiologist review and echocardiographic findings (if referral required).
Results: The study population included 113,694 recruits aged 16 to 19 years which were screened over a 22 year period. Of them, 140 recruits had a history of rheumatic fever (prevalence rate: 0.12%), although none from an Ethiopian background (n = 1,719). The prevalence of valvar disease confirmed echocardiographically in Ethiopian recruits was 0.76% compared with a rate of 0.93% among all recruits ($p = 0.5$). However, the prevalence among Ethiopian recruits was higher ($p < 0.01$) in those who had migrated to Israel aged 12 years or over (2.1%, n = 287), compared to those who migrated at a younger age (0.27%, n = 742) or were born in Israel (0.72%, n = 690) (as a group 0.49%, n = 1432).
Conclusions: These findings suggest that an Ethiopian teenage population from the Gondar region may have had a rate of clinically evident and echocardiographically confirmed RHD of 1.6% (2.1% {prevalence of valvar heart disease assumed to be secondary to RHD + background prevalence}– 0.49% {background prevalence presumed not related to RHD}), high even for Africa despite no apparent past history. The data also implies that this central African population had no genetic predisposition to RHD as those immigrating before 12 years of age or born in Israel had a lower prevalence of valvar disease than the local population.

P2279 - PERICARDITIS IN CHILDREN EXPERIENCE OF A MOROCCAN UNIVERSITY HOSPITAL

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Objective: To describe the profile and the outcome in children with acute pericarditis.
Methods: The medical files of all patients with a diagnosis of acute pericarditis admitted in the pediatrics department in Med VI University Hospital of Marrakech, during January 2015 to November 2016 were analyzed retrospectively for their etiology, presentation, management, and prognosis.
Results: The total number of patients was 8: 6 boys and 2 girls with a mean age of 10.75 (range: 5 years to 14 years). The predominant symptoms were chest pain, tachypnea, and fever. Chest radiograph and echocardiogram were performed in all cases. Cardiomegaly was a constant character in all patients. Echocardiography was performed between the first and twentieth days of hospitalization, 6 children had pericardial effusion, and 2 had cardiac tamponade. The etiology of pericarditis was as follows: purulent or bacterial pericarditis in 4 (50%) cases, tuberculosis in 2 (25%) cases, and rheumatic fever in 2 (25%) cases. 2 patients (25%) underwent pericardiocentesis and 1 child required pericardiotomy. Echocardiographic follow up performed in less than one month showed: 1 case of dry pericarditis, 2 cases of small pericardial effusion and 4 cases were normal. There was no constrictive pericarditis. All patients had a good evolution. However, one patient had an acute pulmonary edema that was resolved. Median duration of evolution towards the disappearance of clinical signs after treatments was 32.5 days (interquartile range 5 to 60 days).
Conclusions: Management of pediatric pericarditis is influenced by the cause of the pericarditis. It may involve supportive care, pain control, and antibiotic therapy, as well as pericardiocentesis or pericardiectomy if demanded. The tuberculosis and rheumatic pericarditis remain frequent in our context.

P2281 - REGIONAL VARIATION IN THE MANAGEMENT OF KAWASAKI DISEASE; PRELIMINARY RESULTS FROM AN ONGOING GLOBAL SURVEY

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Background: Intravenous immunoglobulin (IVIG) significantly reduces coronary artery complications in Kawasaki disease (KD) and are universally accepted for the treatment of KD. However, other interventions are used inconsistently and differences in practice exist across nations and institutions.
Methods: Cross-sectional study on the management of patients with KD using a standardized online survey (SurveyMonkey).
Results: The survey has been completed to date by 188 physicians from 26 countries and 15 pediatric subspecialties. IVIG was used as initial treatment by 91% of physicians. Physicians from countries with lower Human Development Index (HDI) were less likely to treat patients with normal coronary arteries ($p = 0.004$), despite no difference in belief that all patients should be treated ($p = 0.35$). Cost of IVIG and limited availability of immunomodulating therapy were the most common barriers to treatment. Steroids were used more often in Europe and South-America ($p < 0.001$), and anti-TNFα therapies in countries with lower HDI ($p = 0.04$). Risk scores were used by a third of physicians to guide decisions about additional therapies, more often in Asia and Europe ($p = 0.01$). The majority of physicians defined treatment failure as persistent or recrudescent fever 24 hours after the end of IVIG infusion. Persistent fever and increasing coronary artery dimensions were the most important factors in decision to retreat. Treatment of refractory cases consisted of IVIG (82%), steroids (83%) and anti-TNF agents (55%) in the majority of cases. Steroids were used more often in Europe and South-America ($p < 0.001$); and anti-TNFα in Asia and Europe ($p = 0.02$) as well as countries with lower HDI ($p = 0.02$).
Conclusions: Management of patients with KD varies significantly. Medical and economic resources explain some variations in treatment, but country of origin also significantly affects management decisions, irrespective of economic status. Physicians who manage patients with KD are invited to complete the anonymous on-line survey and contribute to this research. https://www.surveymonkey.com/r/internationalKDsurvey.

P2294 - THE EFFECT OF TYPE AND SEVERITY VALVES'S ABNORMALITIES TO NUTRITIONAL STATUS IN CHILDREN WITH RHEUMATIC HEART DISEASES IN SOETOMO HOSPITAL SURABAYA

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Background: Children with rheumatic heart disease most likely demonstrate poor nutrition due to wasting of body fat and skeletal muscle. Evidence for cardiac cachexia maybe seen in different type and severity of valve abnormalities.
Objective: To examined the association between the type and severity of valves abnormalities with nutritional status in RHD children.
Methods: A cross sectional study was done at Paediatric Cardiology Outpatient Clinic of Dr.Soetomo Hospital, Surabaya in period of 2013-2016. Nutritional status was classified into undernourished (underweight and severe underweight), normal and overnourished (overweight and obesity) based on weight for age. Valve abnormalities was studied from echocardiography. Statistical analysis using comparative statistic and a value of $P < 0.05$ considered significant.
Result: A total of 50 children were evaluated [23 (46%) boys; median age 11 (range: 6-15) years]. Nutritional status of undernourished, normal and overnourished were 15 (30%), 27 (54%) and 8 (16%), consecutively. Valve abnormalities at presentation were mitral regurgitation (MR) 43 (86%), aortic regurgitation (AR) 19 (38%) and tricuspid regurgitation (TR) 22 (44%) children. Severity of valves regurgitation were severe MR 22 (44%), severe AR 5 (10%), and severe TR 4 (8%). There were no association between type of valve abnormalities and nutritional status, MR ($P = 0.287$), AR ($P = 0.861$), TR ($P = 0.807$). However, only severity of MR have significant association with nutritional status [MR ($P = 0.022$), AR ($P = 0.628$) and TR ($P = 0.595$)].
Conclusion: Severity of mitral regurgitation associates with nutritional status, but the type of valve abnormalities do not associate with nutritional status.

P2305 - REACTIVATION OF RHEUMATIC FEVER IN RHEUMATIC HEART DISEASE IN INDONESIA COMPARISON BETWEEN CHILDREN AND YOUNG ADULTS

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Background: Rheumatic heart disease (RHD) remains one of the largest preventable burdens of disease in the world. Whereas repeated episodes of rheumatic fever (RF) become an important mechanism for worsening of RHD. The proportion of the disease remains high in the developing countries, associated with lack of access to medical facilities, infrastructure and education for preventive programs.
Objectives: This study aimed to evaluate the pattern of RF reactivation in in the established RHD among Indonesian children and young adult.
Methods: This retrospective study was conducted at National Cardiovascular Center Harapan Kita, Jakarta, as the tertiary cardiovascular center in Indonesia from January 2012 to December 2015. RF reactivation was based on revised Jones criteria and 2002-2003 WHO criteria.
Results: Among patients with RHD, there were 84 children with median age of 12.5(2-17) years, 103 young adults with median age of 24(18-30) years, 88(47.0%) male and 99(52.9%) female. Reactivation of RF was found in 39(20.8%), and significantly higher in children 28(33.3%) than in young adults 11(10.6%), $p < 0.001$. The number of patient treated with phenoxy methyl penicillin as secondary prevention was only 51(27.2%) patients, where 34 (40.4%) were children and 17(16.5%) were young adults ($p < 0.001$). Twenty five (13.4%) patients were hospitalized with severe congestive heart failure and 42(22.5%) patients with pulmonary hypertension.
Conclusion: The number of cases with reactivation of RF was higher in the children age group and the coverage of secondary prevention was still low. Secondary prevention should be more encouraged since it remains the most practical solution to reduce the worsening of RHD in the developing countries.

P2310 - CLINICAL PROFILE RHEUMATIC HEART DISEASE AT SOETOMO HOSPITAL SURABAYA

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Background: Rheumatic Heart Disease (RHD) affected 19 per 100.000 children in developing countries. There are only few

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systematically collected data on diseases characteristic in Dr. Soetomo Hospital, Surabaya.
Objective: To describe the clinical profile of pediatric patient with RHD.
Methods: Demographic data, and clinical details regarding RHD in pediatric cardiology outpatients clinic Dr. Soetomo Surabaya Hospital from June 2014 to June 2016 were collected. Missing information obtained directly from the patients' parents or guardians.
Results: Among total of 50 patients, majority were school aged (6-11 years old) with median age 10.86 years (ranged 6–15 years). Female to male ratio was 1.17 :1. Most patients lived outside Surabaya 27 patients (54%), 33 patients (66%) were referral patients. At the first time of diagnosis, there were 14 patients (28%) with ARF (Acute Rheumatic Fever) and 36 patients (72%) with RHD. Major clinical manifestations were carditis 13 patients (26%), followed by polyarthritis migrains 1 patient(2%) and chorea 1 patient(2%). Minor clinical manifestations were fever 7 patients (14%), arthralgia 2 patients(4%), positive CRP 30 patients(60%), elevated ESR 34 patients(68%). Supportive evident positive ASTO titer 22 patients(44%). From electrocardiography findings, only 1 patient had prolonged PR interval, and 2 patients had axis deviation. The most common valve lesions were severe MR 20 patients(40%) followed by moderate MR 17 patients(34%). Most of them 27 patients(54%) had normal nutritional status.
Conclusions: Carditis was the most common major clinical manifestation in previous ARF and mitral regurgitation was the most common valve lesion in patient with RHD.

P2371 - A SYSTEMATIC REVIEW OF THE PREVALENCE OF GROUP A STREPTOCOCCAL DISEASE IN AFRICA

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Introduction: The prevalence of group A streptococcal (GAS) disease is estimated at >18.1 million cases with an incidence of >1.78 million cases py. While GAS is a significant cause of mortality and morbidity on the global scale, the burden of GAS disease in Africa is not known.
Methods: We conducted a comprehensive literature search in a number of databases, using an African search filter to identify published studies of GAS prevalence amongst symptomatic patients. Two reviewers independently selected articles meeting pre-specified criteria and extracted data on a standardised data extraction form. Statistical analysis included the identification of data sources, documenting estimates, and the application of the random-effects meta-analysis model to aggregate prevalence estimates 95% CI for GAS. We applied Freeman-Tukey transformation to account for between study variability. The I^2 statistic was used to determine the extent of variation in effect estimates due to heterogeneity (H0).
Results: Of 128 articles identified, 27 met the inclusion criteria. Meta-analysis revealed a pooled prevalence of 23% (95% CI, 20–27%). Regional pooled prevalences were similar between Western, 25% (95% CI, 23–27%) and Northern Africa, 26% (95% CI, 21–31%). Central and Eastern Africa had the lowest pooled estimate of 5% (95% CI, 2–11%) and 16% (95% CI, 8–27%) respectively, while Southern Africa had the highest pooled estimate of 32% (95% CI, 20– 46%). Two articles reported on molecular characterisation of isolates with 92 *emm*-types documented; the most dominant subtypes were *emm*44, 48 and 65.
Conclusion: The prevalence of GAS is high in Africa. Efforts focused on the primary prevention of GAS pharyngitis are therefore warranted. Data on molecular strain characterization of GAS in Africa is largely non-existent and thus there is a need for further studies such as AFROStrep to inform current prevention efforts including vaccine development.

P2403 - THE NEW ZEALAND FAMILIAL ECHO STUDY REVEALS HIGH PREVALENCE OF RHEUMATIC HEART DISEASE AMONGST PARENTS AND SIBLINGS OF CHILDREN WITH RHEUMATIC FEVER ECHOCARDIOGRAPHY SHOULD BE OFFERED TO FIRST DEGREE RELATIVES

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Background: The risk of Rheumatic Heart Disease (RHD) in relatives of Acute Rheumatic Fever (ARF) patients is poorly defined. This study aimed to determine the prevalence of RHD in first degree relatives of ARF patients using echocardiography and family history, and to explore the feasibility of using echocardiography for enhanced case detection.
Methods: Between 2014 – 2016, 70 cases of ARF were recruited from Auckland, New Zealand (NZ). Family composition and family history of ARF/RHD were determined via structured questionnaire. Parents and siblings ≥4 years were offered echocardiography. Echocardiograms were interpreted according to WHF 2012 criteria. Abnormal scans were read by a panel of 3 cardiologists.
Results: 70 cases and families were recruited. The median age of cases was 11 years (range 4 – 15 years). 55/70 (79%) were of Pacific Island ethnicity and 15/70 (21%) were New Zealand Maori. 132/166 (80%) of eligible siblings and 94 parents underwent echocardiography. Siblings: Echocardiography detected 2 with Definite RHD and 7 with Borderline RHD. 1 additional sibling had known RHD following an ARF episode. Parents: Echocardiography detected 3 with Definite RHD and 3 with Borderline RHD. 2 additional parents had known prior RHD. Prevalence of RHD for parents and siblings is shown in Table One. Overall, the prevalence of RHD (Definite + Borderline) in siblings was 75 per 1,000 (95% CI 40 – 134 per 1,000) and in parents was 83 per 1000, 95% CI 41 – 158 per 1,000.).
Conclusions: We report 8% prevalence of RHD in parents and siblings of ARF cases in New Zealand. These findings support the concept that susceptibility to ARF has a heritable component. In adequately resourced healthcare systems, first degree relatives should be offered echocardiography when a family member is diagnosed with ARF.

Table 1. Prevalence of RHD in parents and siblings of 70 children with ARF

	Borderline RHD		Definite RHD		Total RHD (Definite + Borderline)	
	Rate	Prevalence	Rate	Prevalence	Prevalence	95% CI
Siblings	7/133	53 per 1000	3/133	23 per 1000	75 per 1000	40–134 per 1000
Parents	3/96	31 per 1000	5/96	52 per 1000	83 per 1000	41–158 per 1000

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P2470 - PREVALENCE OF POSITIVE RAPID ANTIGEN GROUP A STREPTOCOCCUS TEST IN CHILDREN AND ADOLESCENTS IN A STATE FROM NORTHEAST BRAZIL

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Background: Group A streptococcus (GAS) is deeply associated with sore throat that can lead to acute rheumatic fever and rheumatic carditis. Prevalence of asymptomatic carriers (AC) in Brazil has been reported from 0.8–1.6%. This statistic may vary with the quality of health care provided. Antibiotic therapy with penicillin is effective and low cost, but in 2015 and 2016, Brazil faced shortage of the drug caused by global raw material destocking. This study aims to determine the prevalence of AC through rapid antigen group A streptococcus test (RAGAST) in a poor state in Northeast of Brazil.
Materials and Methods: Analysis of data of 1,648 children over age four and adolescents evaluated in the Heart Caravans, an active search for heart disease that takes place once a year in 13 cities of state of Paraiba, in northeast of Brazil. All patients were submitted to RAGAST using Quidel QuickVue+ kit. Prevalence data was analyzed each year and compared with other variables such as age, social and economic status and diagnosis of rheumatic fever.
Results: We found a high prevalence of positive tests each year, varying from 14.3% in 2014, to 4.3% in 2015 and 21.9% in 2016 ($p < 0.01$). During this time, variables associated with higher prevalence were male gender (p 0.032), age (p 0.034), brown ethnicity (p 0.041) and access to public water supply system (p 0.018).
Conclusions: A high prevalence of GAS AC was demonstrated in the studied population. A reduction in prevalence was observed between 2014 and 2015, with a greater increase between 2015 and 2016. We theorize that the lack of appropriate antibiotic therapy between 2015 and 2016 may explain such a rise. The Heart Caravan intervention in 2014 may have contributed in decreasing prevalence between 2014 and 2015.

P2534 - RISK FACTORS AND IMPLICATIONS OF PROGRESSIVE CORONARY DILATATION IN CHILDREN WITH KAWASAKI DISEASE

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Background: Kawasaki disease (KD) is an acute systemic vasculitis of childhood, which may lead to cardiovascular morbidity and mortality. Progressive coronary dilatation at least two months has been known associated with worse late coronary outcomes in the KD patients with medium or giant aneurysms. However, its risk factors and occurrence in the KD patients without medium or giant aneurysms were less studied.
Method: We retrospectively enrolled 169 KD patients from a tertiary medical center (2009-2013). Medical records were reviewed. Echocardiography was followed at the acute KD phase and 3-4 weeks, 6-8 weeks, 6 months and 12 months after disease onset. Progressive coronary dilatation was defined for those with progressive enlargement of coronary arteries in three consecutive echocardiograms.
Results: Maximal coronary Z-score of 31 KD patients (31/169, 18.3%) were more than +2.5 at the acute phase KD. 16 patients (16/169, 9.5%; M/F:9/7) had coronary aneurysms one month after KD onset and 5 (5/169, 3.0%) of them satisfied the definition of progressive coronary dilatation. By using multivariate logistic regression, we found initial maximal coronary Z-score $\geqq +2.5$ [odds ratio (OR): 5.24, $P = 0.020$] and hypoalbuminemia [OR: 4.83, $P = 0.035$] were two independent risk factors of coronary aneurysms. Both factors were also significantly associated with progressive coronary dilatation. IVIG unresponsiveness was borderline associated only with the development of coronary aneurysms one month after disease onset ($P = 0.058$).
Conclusions: 3% of KD patients had progressive coronary dilatation, which was associated with the aneurysm persistence one year after KD onset. Initial coronary dilatation and hypoalbuminemia were independently associated with the occurrence of progressive coronary dilatation. Such patients may need closer cardiac monitoring and adjuvant therapies besides immunoglobulin.

P2540 - PATTERN OF RHEUMATIC HEART DISEASE AMONG CHILDREN YOUNG ADULTS AND ADULTS IN INDONESIA

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Background: Rheumatic heart disease (RHD) continues to be a major public health problem in the developing countries. The aim of this study was to investigate the pattern of RHD among Indonesian children, young adults, and adults whom hospitalized at the tertiary center in Indonesia.
Methods: This retrospective study was conducted at National Cardiovascular Center Harapan Kita, Jakarta, Indonesia. The data were retrieved from the hospital database in one year period.
Results: Total of 64 patients, 34.4% males and 65.6% females were included in this study. Patients were divided into 3 groups, 21 (32.8%) children (<18 years old), 18 (28.1%) young adults (18-30 years old), and 25(39.1%) adults (>30 years old). The most common clinical presentation was congestive heart failure found in 66.7% children, 88.9% young adults, and 13(5%) adults. There was not significantly different in this clinical presentation between groups ($p = 0.05$). The most valve disease findings among children group were isolated mitral regurgitation (85.7%), followed by both mitral and aortic regurgitation (9.5%). There were 72.2% patients with isolated mitral regurgitation and 16.7% with isolated mitral stenosis in young adults group. While in adults group, 68% with mitral stenosis, 16% patients with combination of mitral stenosis and regurgitation. These valve disease findings was significantly different between groups ($p = 0.000$). Among children group, there was 47.6% patients only treated medically and 33.3% whom underwent surgical mitral valve repair. In young adults, mitral valve replacement and mitral valve repair performed in 33.3% each. Medical treatment only, mitral valve replacement and mitral valve repair were done in 32%, 40%, and 20% respectively in adults group. These procedures were also not significantly different between groups ($p = 0.051$).
Conclusions: The RHD patterns in our institution, apart from type of valve involvement, there was no difference of clinical presentation and method of treatment between children, young adults, and adult patients.

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P2682 - TWO YEAR FOLLOW UP OF RHEUMATIC HEART DISEASE DIAGNOSED THROUGH ECHOCARDIOGRAPHIC SCREENING IN MALAWI AFRICA

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Background: In asymptomatic children, screening echocardiography (echo) may diagnose latent rheumatic heart disease (RHD). World Heart Federation has standardized categorization of "definite", "borderline", or no RHD by echo findings. Progression of screening-diagnosed RHD is not known. In 2014, we screened 1450 schoolchildren in Lilongwe, Malawi and diagnosed 39 with borderline and 11 with definite RHD.
Objective: Evaluate 2-year RHD evolution amongst those diagnosed through screening.
Methods: 2-year follow-up echos were performed on participants with definite and borderline RHD. Echos were read by primary, secondary, then third readers if there was disagreement. We compared initial and follow-up diagnosis and compared penicillin adherence, age, gender, number in home, and household income among those with disease advancement versus stable disease. Comparisons used Fisher's exact and Wilcoxon rank sum tests. If children did not come to follow-up appointments, they were traced using location information provided at enrollment.
Results: Of 39 with borderline RHD, 1 was lost to follow-up (2.6%), 1 progressed to definite (2.6%), 19 remained borderline (48.7%), 17 (43.6%) regressed to normal, and 1 was re-classified as mitral valve prolapse (2.6%). Of 11 with definite RHD, 6 (54.5%) remained definite, 4 regressed to borderline (36.4%), and 1 regressed to normal (9.1%). Only 5 of 11 (45%) with definite RHD had penicillin adherence >80% for the previous year. There was no difference in adherence, gender, age, household income, or number in household between those with definite RHD that regressed and those who did not ($p > 0.05$), or between those with borderline who regressed and those that did not ($p > 0.05$). Retention rate was 98%.
Conclusions: A strength of our study was a high retention rate. At 2 years, more patients had disease regression than progression similar to previous cohorts, but longer follow-up is required to predict true disease evolution.

P2764 - EFFECTIVENESS OF A NURSE LED MECHANICAL VALVE ANTICOAGULATION PROGRAM FOR RHEUMATIC HEART DISEASE PATIENTS IN HAITI

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Introduction: Rheumatic heart disease (RHD) disproportionately affects young people in low- and middle-income countries (LMIC) where health systems for chronic care are weak. Mechanical valves are more durable than bioprosthetic valves, but require long-term anticoagulation. In LMICs, bioprosthetic valves are often chosen given concerns about the feasibility of long-term anticoagulation.
Hypothesis: Nurse-led management allows effective anticoagulation in Haiti.
Methods: A nurse-led anticoagulation program was established in Haiti. 25 RHD patients with mechanical valves (7 female, median age 19 years) were followed from 9/2013 through 12/2016. All patients live in impoverished communities: 15 urban, 10 rural. Valve position was mitral only (17), aortic only (3), or mitral and aortic (5). We trained patients to use donated portable machines (Coagusense Inc., Fremont, CA) to measure their International Normalized Ratio (INR). On a defined schedule, their INR values were checked and self-reported by phone to supervising nurses. Warfarin dose was titrated by standardized protocol. Physician support was available. Time in therapeutic range (TTR) was calculated using the modified Rosendaal method.
Results: In 907 INR samples, median TTR was 50.1% (Interquartile Range [IQR] 30.6% - 61.7%) (Table 1). Time below and above therapeutic range was 35.2% (IQR 27.9% - 46.8%) and 14.7%, (IQR 7.1% - 20.9%), respectively. INR was within 1.0 of target 95.0% (IQR 94.1% - 98.6%) of the time. One patient died unrelated to anticoagulation due to severe dilated cardiomyopathy. Twenty-four participants reported no anticoagulation-related complications (severe bleeding, thromboembolism, endocarditis, or valve dysfunction). INR test results were reported to the nurse a mean of 4.3 days after the protocol-specified date.
Conclusion: Nurse-led, protocol-driven anticoagulation of patients with mechanical valves can be successfully and safely managed in a low-income country. Health systems including physician-nurse task shifting can be implemented to facilitate long-term care of patients with RHD in whom surgery with mechanical valve replacement is required.

Table 1.

Patient #	Age	Sex	Valve Position	Low INR Limit	High INR Limit	Months in Program	Total Tests	% of Days In Range	% of Days Out Of Range (1.0 or less)	% of Days Out Of Range (more than 1.0)
1	22	M	Mitral	2.5	3.5	39	107	44.3%	50.5%	5.2%
2	20	M	Mitral/ Aortic	2.5	3.5	39	54	51.3%	46.7%	2.0%
3	18	M	Mitral	2.5	3.5	33	61	59.7%	37.1%	3.2%
4	24	M	Mitral/ Aortic	2.5	3.5	35	61	69.8%	23.4%	6.8%
5	22	F	Mitral	2.5	3.5	34	53	28.8%	63.3%	7.8%
6	22	F	Mitral/ Aortic	2.5	3.5	38	44	72.1%	24.0%	3.9%
8	28	M	Mitral	2.5	3.5	33	43	55.3%	38.8%	5.9%
9	18	M	Mitral/ Aortic	2.5	3.5	31	73	30.6%	62.0%	7.4%
10	14	F	Mitral	2.5	3.5	3	11	27.5%	60.6%	11.9%
11	17	M	Mitral	2.5	3.5	24	46	62.7%	35.2%	2.0%
12	11	M	Mitral	2.5	3.5	21	31	45.1%	54.7%	0.2%
13	22	F	Mitral	2.5	3.5	18	43	53.2%	44.5%	2.3%
14	18 at Death	M	Mitral	2.5	3.5	10	24	1.8%	64.3%	33.8%
15	31	M	Mitral/ Aortic	2.5	3.5	15	33	54.0%	43.0%	3.0%
16	19	M	Mitral	2.5	3.5	13	26	68.0%	30.2%	1.8%
17	24	M	Aortic	2	3	12	28	64.9%	34.4%	0.8%
18	22	F	Mitral	2.5	3.5	12	22	53.7%	46.3%	0.0%
19	24	F	Mitral	3	3.5	12	45	38.9%	57.1%	4.0%
20	21	M	Aortic	2.5	3.5	9	24	17.0%	81.6%	1.4%
21	19	M	Mitral	2.5	3.5	8	28	60.6%	35.5%	3.9%
22	10	M	Mitral	2.5	3.5	7	9	2.3%	97.7%	0.0%
23	10	F	Aortic	2.5	3.5	7	18	48.4%	51.6%	0.0%

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Table 1. *Continued*

Patient #	Age	Sex	Valve Position	Low INR Limit	High INR Limit	Months in Program	Total Tests	% of Days In Range	% of Days Out Of Range (1.0 or less)	% of Days Out Of Range (more than 1.0)
24	16	M	Mitral	2.5	3.5	6	8	65.9%	7.9%	26.2%
25	19	M	Mitral	2.5	3	4	12	34.9%	59.7%	5.4%
26	18	M	Mitral	2.5	3.5	1	3	28.6%	71.4%	0.0%
Total						**464**	**907**	**50.1%**	**46.7%**	**5.0%**
Median	**19**					**13**	**31**	**51.3%**	**46.7%**	**3.2%**
25% Quartile						**8**	**22**	**30.6%**	**35.5%**	**1.4%**
75% Quartile						**33**	**50**	**61.7%**	**61.3%**	**6.4%**

P2837 - CARDIAC ABNORMALITIES IN CHILDREN WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Systemic lupus erythematosus (SLE) is an autoimmune disease characterised by disturbances in innate and adaptive immune mechanisms. Manifestations affecting the skin, joints, kidney and cytopenia are commonly detected in this disease. However cardiac involvements are often subclinical and then misdiagnosed unless they results in severe disorders that could be life-threatning.
Objective: The aim of the study was to assess the systolic and diastolic function of the left ventricle (LV) in SLE children without clinically evident cardiovascular disease, using pulsed Doppler echocardiography.
Methods: We conducted a transversal study in the department of Pediatrics of Sahloul hospital-Sousse. Children and adolescents with SLE and no cardiac symptoms were included in our study. They were examined for cardiac involvement by physical examination and echocardiography.
Results: We collected 4 cases of SLE (2girls, 2 boys). Echocardiographic examination revealed mild tricuspid valve regurgitation in all patients, and moderate mitral valve regurgitation in one patient. Pulmonary arterial hypertension was found in one case (systolic pulmonary hypertension = 30 mmHg). Pericardial effusion was found in one patient. Indexes of LV systolic were correct with ejection fraction (EF) between 59% and 68% and fractional shortening (FS) between 33% and 46%. Peak early diastolic filling velocity (E) was >1,2 m/s in 2 cases and ratio of early-to-late diastolic filling velocity (E/A) was >1 in all cases and >2 in 2 cases. Deceleration time (DT) was long in 2 patients (243ms, 239ms) and low in one case (136ms). Late filling velocity (A) and isovolumic relaxation time (IRT) were low (>60ms) in 2 cases. Patients demonstrated significantly high E'/A' (>1) in all cases.
Conclusion: We conclude that asymptomatic diastolic dysfunction is common in children with SLE, most likely representing myocardial involvement. Routine cardiac evaluation by echocardiography can be recommended in the follow-up of children with SLE in order to detect silent cardiac abnormalities.

P3032 - LONG TERM OUTCOME OF GIANT CORONARY ARTERY ANEURYSM IN KAWASAKI DISEASE

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Purpose: In childhood Kawasaki disease (KD), giant coronary artery aneurysm (CAA) is associated with significant mortality and morbidity associated related to ischemic heart disease. The aim of this study was to investigate the incidence, treatment modalities and clinical course of ischemic cardiac sequelae in KD patients with giant CAA.
Methods and Results: The medical records of children with KD with giant CAA admitted to Chungnam National University Hospital, Konyang University Hospital and Soonchunhyang University Cheonan Hospital from 1993 to June 2016 were retrospectively analyzed. Giant CAA defined as internal diameter >8mm or Z score of coronary artery diameter >10. The subject included 22 patients, who male patients were predominant (90.9%). One patient diagnosed as KD with giant CAA by chance during the evaluation of cardiac murmur. The mean age at diagnosis was 38 ± 32 months and the average follow-up period was 7.4 ± 7.6 years. Four patients (18%) were treated with thrombolytic therapy for acute coronary syndrome at 1 month to 21 months after KD onset and one of them died. The catheter or surgical coronary artery intervention to alleviated coronary ischemia were performed at 1 year to 16 years after onset in three patients (13.6%). The overall all ischemia event free survival rates were 78.6%, 69.9% and 46.6% at 5, 10 and 20 years after the onset of KD, respectively. The age and coronary artery diameter at diagnosis were related to ischemic events but it was not statistically significant.
Conclusion: KD patients with giant CAA were suffered form significant myocardial ischemia in long-term follow up. Age and initial diameter of CAA are related to myocardial ischemia but not significant. The treatment modality including warfarin and antiplatelet therapy was not directly associated with myocardial ischemia. The national multicenter and prospective studies would be suggested in future.

SURGERY

P1003 - COMPARISON WITH TWO TYPES OF PALLIATIVE PROCEDURES FOR TETRALOGY OF FALLOT THE MODIFIED BLALOCK TAUSSIG SHUNT AND THE RIGHT VENTRICULAR OUTFLOW PATCH

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Background: When infants with TOF/pulmonary atresia or low body weight not amenable to initial complete correction, palliative procedure will be performed first. Systemic or right ventricular to pulmonary connection can be established with Modified Blalock–Taussig shunt (MBTS) or surgical right ventricular outflow tract (RVOT) patch enlargement.
Methods: A 7-year retrospective study of TOF cases under 6 month old with palliative procedure without initial total correction in our institute between March 2007 to April 2015 was conducted. Cases were classified into two groups; group I included patients that had mBT shunt o central shunt, while group II included cases that had transannular RVOT patch augmentation. We compared age and body weight at the time of palliative surgery, age at the time of total correction, numbers of operations between palliation and total correction, and pulmonary growing rate of two groups.
Results: Thirty-six patients fulfilled the study criteria. There were 29 cases (80%) in group I, and 7 cases (20%) in group II. All patient tolerated surgical repair but 4 patient expired after operation due

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to sepsis, heart failure, hypoxic-ischemic encephalopathy. Median weight at the time of palliative surgery($p = 0.007$) in group I and group II is 2.9 kg(mean, 3.13 ± 0.96; range, 1.8 to 6.2 kg) and 2.3 kg(mean, 2.25 ± 0.26; range, 1.7 to 2.5 kg). Numbers of operations between palliation and total correction in group II is zero. In group I, 9 of 29 patients had one time operation between palliation and total correction. We observed no statistically significant difference in pulmonary artery growing rate (mm2/days).
Conclusions: Transannular RVOT patch augmentation with cardiopulmonary bypass can be performed safely and effectively in lower age and body weight infants as palliative procedure. It also can decrease the number of operations between palliation and total correction. Long-term follow up is needed to evaluate future difference in different techniques.

P1009 - IMPACT OF SOCIAL DISADVANTAGE ON LONG TERM OUTCOMES IN CHILDREN UNDERGOING CARDIAC SURGERY

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Background: Population based registries report 95% 5-year survival in children undergoing surgery for congenital heart disease. The Australian indigenous population is at risk from social disadvantage, inequitable access to healthcare and inadequate follow-up. This study investigates paediatric outcomes and influencing factors in this at risk population.
Methods: All children with acquired or congenital heart disease who underwent cardiac surgery between May 2008 and August 2014 were studied. Demographic information, diagnosis and comorbidities, treatment and outcome data were collected at time of surgery and at cardiac follow-up. Socio-economic Indexes for Areas-2001 was used to quantify socioeconomic status of patients.
Results: 1528 children with mean age 3.4 ± 4.6 years were studied. 123 (8.1%) were identified as indigenous patients. Assessment of socioeconomic status demonstrated that 64 (52.7%) indigenous patients were in the lowest third of the population, compared to 456 (28.2%) of the non-indigenous population ($p = < 0.001$). There was no difference in the Basic Aristotle scores between populations ($p = 0.20$) The indigenous population had a significantly higher Complex Aristotle score (indigenous 9.4 ± 4.2 vs non-indigenous 8.7 ± 3.9, $p = 0.04$). Rate of long-term follow-up did not differ between groups (indigenous 93.8% vs. non-indigenous 95.6%, $p = 0.17$). No difference was noted in 30 day mortality (indigenous 3.2% vs non-indigenous 1.4%, $p = 0.13$). 6-year survival for the entire cohort was 94.9% with cox analysis demonstrating increased 6-year mortality in the indigenous group (indigenous 8.1%% vs non-indigenous 5.0%; hazard ratio = 2.10; $p = 0.03$). Freedom from surgical re-intervention was 79.5% for all patients and did not differ between groups (hazard ratio = 1.40; $p = 0.11$). When long-term survival was adjusted for socio-economic status and Complex Aristotle Score, no significant difference in outcomes between the populations was demonstrated ($p = 0.19$).
Conclusion: Despite equitable access to cardiac medical care, the indigenous population experienced a higher late mortality, which may be explained by complex socioeconomic issues.

P1021 - CASE REPORT AORTO VENTRICULAR TUNNEL

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One day old male, product of a 39 week uncomplicated gestation, labor and, delivery to 19 yr.old female was noted to have respiratory distress characterized by intercostal retractions and nasal flaring shortly after birth. On physical assessment, a systolic murmur, III/IV and an intense second heart sound, were detected. Chest x-ray demonstrated cardiomegaly ; echocardiogram revealed an atrial communication with a left to right shunt, a ductus arteriosus with bidirectional flow, and a direct communication between the left ventricle (LV) and a dilated right sinus of Valsalva. There was anterograde flow into the aorta in systole and retrograde flow into the ventricle in diastole. Despite treatment with diuretics, the baby developed refractory cardiac failure. The baby was scheduled for surgery. The procedure was performed with aortic cross-clamping, cardioplegia, ligature of both cavae and a right atriotomy. We observed a tissue like bump over the right sinus of valsalva to the anterior left ventricular outflow tract (LVOT). We opened this tissue and found a communication between Aorta and LV, Aortic valve was complete and without problems, right coronary artery was normal size; and then it was closed along its anterior wall with a Goretex patch using a double running suture; same patch was used to close LVOT, use running suture over aortic annulus and close the Aorta. We close the remnant tissue over the patch. The ASD and the atriotomy were repaired. The aortic clamp and caval ligatures were released. Heart rate returned to sinus rhythm with the aid of dobutamine, epinephrine and milrinone. Cardiopulmonary bypass time was 55 min. and the aortic cross-clamping, 43 min. Post-op management included iv fentanyl, mechanical ventilation and continued administration of inotropic drugs. Over the second day post-op the baby.

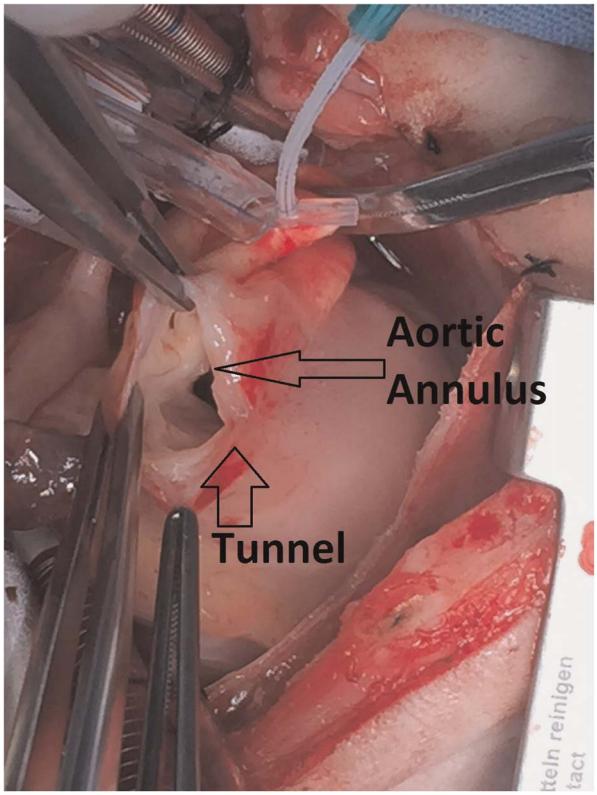

Figure 1.

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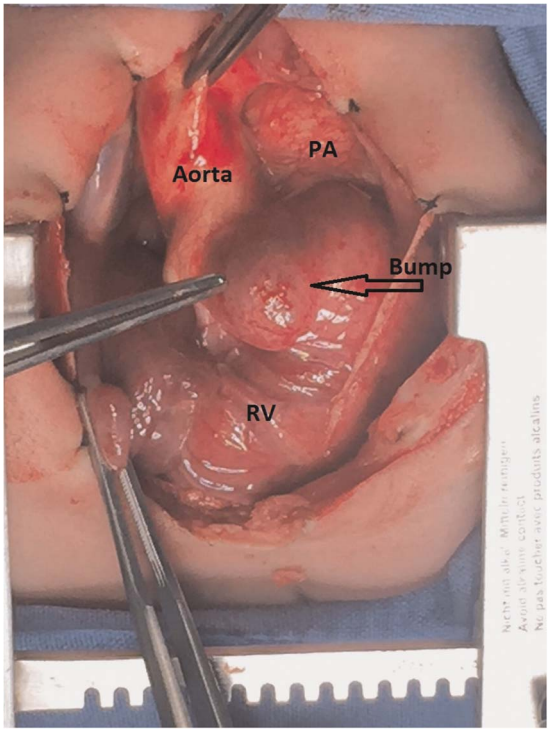

Figure 2.

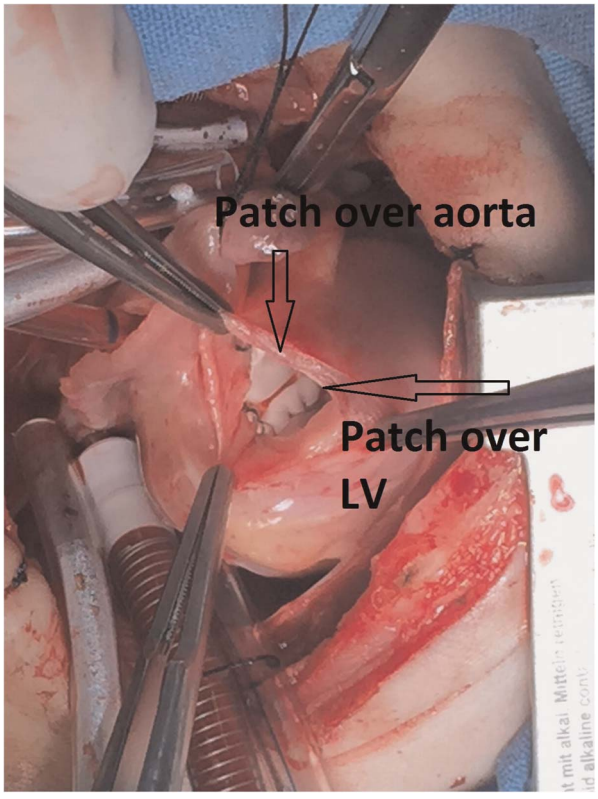

Figure 3.

P1024 - MITRAL VALVE REPLACEMENT FOR A PATIENT LESS THAN TWO YEARS OF AGE WITH MITRAL REGURGITATION AND EVOLVING POST REPAIR MITRAL STENOSIS

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We present a case of a one year and nine-month old girl who was diagnosed by echocardiography to have congenital mitral regurgitation at four months of age after presenting with a murmur and heart failure symptoms. Despite being maintained on medications, the mitral regurgitation became progressively worse, associated with rapid enlargement of the left-sided chambers, prompting mitral valve repair at 14 months old. Although intra-operative transesophageal echocardiography showed only moderate mitral regurgitation, serial echocardiographic studies done on the 3rd month and 6th month post-repair showed rapidly evolving mitral stenosis, associated with recurrence of heart failure symptoms. She therefore underwent mitral valve replacement, seven months after the first repair, using a 19mm bioprosthetic aortic valve. This case report highlights the surgical aspects of repairing a congenitally incompetent mitral valve, and the challenging dilemma of choosing between mitral valve repair versus replacement in the very young pediatric patient. Key words: Congenital mitral regurgitation, Mitral valve Repair, Mitral Valve Replacement.

P1035 - NEONATAL CARDIAC LUPUS MYOCARDITIS FLAIL TRICUSPID VALVE AND NORMAL RHYTHM

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Background: Neonatal cardiac lupus is a rare, passively acquired autoinmune disease. Congenital atrioventricular block is the most common cardiac abnormality found. We describe the first case of flail tricuspid valve due to papillary muscle rupture following in utero myocarditis associated with maternal anti-Ro and anti-La antibodies in absence of atrioventricular block. We discuss the pathophysiology and management in this unique scenario.
Case: We present a patient who was first detected in utero myocarditis. The mother was identified as antibody positive for lupus (anti-SSA/Ro and anti-SSB-La). Dexametasone was started. Fetal echocardiography showed signs of acute myocarditis, with involving of both the chordal and subvalvar apparatus. Normal rhythm was maintained. At 34th week, an emergent caesarean was performed. Following birth, echocardiography showed a flail tricuspid valve due to anterior papillary muscle rupture and severe tricuspid regurgitation. Treatment was initiated (dopamine, milrinone, hydrocortisone and immunoglobulin). Anti-SSA/Ro and anti-SSB-La were positive in the neonate. He was extubated on the 5th day after birth. Symptoms of heart failure improved (tricuspid regurgitation unchanged). He was discharged at home on day 25 after birth on medical treatment. At 2 months of age (weight 3.6 kg), he underwent tricuspid valve repair (7/0 polytetrafluoroethylene neochordae was placed onto the anterior leaflet with attachment to the papillary muscle; no associated annuloplasty; total cardiopulmonary bypass 46 minutes; aortic cross-clamping time 23 minutes).
Results: No postoperative complications were detected. He was discharged on the 26 postoperative day. At 20-month follow-up,

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he is progressing well, with normal-sized right ventricle and mild tricuspid regurgitation.
Conclusions: The spectrum of cardiac manifestations associated with anti-SSA/Ro and SSB/La antibodies is expanding. The recognition of flail tricuspid valve as the final event of an underlying myocardial auto-immune inflammatory process may be added as manifestation of neonatal lupus. The management of heart failure in the neonate is mandatory for a proper timing for surgery.

P1037 - TRAINING ANIMAL MODEL PROGRAM FOR VIDEO ASSISTED REPAIR OF CONGENITAL HEART DEFECTS IN PEDIATRIC PATIENTS INITIAL EXPERIENCE

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Background: Minimally invasive cardiac surgery has grown in adult population. However, its application to congenital heart disease in pediatric patients remains limited. In this scenario, alternative minimally invasive approaches have recently been used, but taking a step forward on a video-assisted repair is still unclear. We present the initial experience of a pioneering training animal model program for video-assisted repair in congenital heart defects in pediatric patients. We discuss the preliminary results in this complex scenario.
Patients and Methods: Phase 1. The ovine model (total number: 8) was used for a progressive training of the pediatric cardiac surgeons under the supervision of expertise pediatric surgeons in videothoracoscopy. Training areas: thorax access (three working ports), dissection and handling of great vessels and adjacent structures (in vivo); right atriotomy, creation of atrial septal defects (ASD) and direct closure (post-mortem). Material: camera (0° y 30°-angled; KarlStorz material), trocars (5-11 mm) and additional surgical material. Phase 2. Patients. Initially, 4 pediatric pacientes (table) with ASD underwent minimally invasive video-assisted repair (submammary mini-thoracotomy, two working ports and peripheral cannulation).
Results: Direct closure was performed uneventfully in all them. Intraoperative extubation was performed in 3. One significative postoperative complication was presented in the patient #4 (reperfusion injury of right lower limb secondary to cannulation, resolved with decompressive fasciotomy). At follow-up (11 + /-2 months, range 8-13), all remain with optimal functional class with no residual defects.
Conclusions: Although the small size of pediatric patients constitutes the main limiting factor for a video-assisted repair, the use of animal models for training in these techniques may be useful. This training in close partnership with expertise pediatric surgeons in thoracoscopic techniques is a key point for acquiring novel surgical skills for those surgeons dedicated to congenital heart defects, improving the presented preliminary results in pediatric patients in a future..

Table 1. Anthropometric and surgical features of the pediatric patients.

	Gender	Age (years)	Weight (kg)	CPB time	AO-Cross clamping time
#1	Male	12	36	145	70
#2	Male	13	46	157	41
#3	Male	12	23	102	23
#4	Female	10	50	135	52

CPB: Cardiopulmonary bypass time.

P1039 - MULTICENTER QUALITY IMPROVEMENT PROJECT TO PREVENT STERNAL WOUND INFECTIONS IN PEDIATRIC CARDIAC SURGERY PATIENTS

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Background: Children undergoing cardiac surgery are at risk for sternal wound infections (SWI) leading to increased morbidity and mortality. Single center quality improvement initiatives have demonstrated decreased infection rates utilizing a bundled approach. The project's purpose was to determine the efficacy of a protocolized approach to decrease SWI.
Methods: Pediatric cardiac programs joined a collaborative effort to prevent SWI. Programs implemented the protocol, collected compliance data and provided data points from local clinical registries using Society of Thoracic Surgery Congenital Heart Surgery Database harvest-compliant software or from other registries.
Results: Nine programs prospectively collected compliance data on 4198 children. Days between infections were extended from 68.2 days (range 25-82) to 130 days (range: 43-412). Compliance was 76.7% the first quarter and 91.3% the final quarter. Ninety (1.9%) children developed a SWI pre protocol and 64 (1.5%) post protocol, $p = 0.18$. The 657 (15%) delayed sternal closure (DSC) patients had a 5% infection rate with 18 (5.7%) year one and 14 (4.3%) year two. For DSC patients there was a trend towards increased risk for SWI by 1.046 for each day the sternum remained open, $p = 0.067$. Single dose bolus pre-op antibiotics administered 0- 60 minutes pre incision resulted in less infections than when administered outside 0-60 minutes, 3.7% v. 1.9%, $p = 0.025$.
Conclusions: A multicenter QI project to reduce pediatric SWI's demonstrated an extension of days between infections and a decrease in SWI's. Administration of pre-op antibiotics within the recommended time was associated with decreased infection rates.

P1055 - A COMPLICATED CASE OF INFECTIVE ENDOCARDITIS COMPLICATING COMPLETE HEART BLOCK POST SURGERY A CASE REPORT

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Introduction: Infective endocarditis (IE) is a serious infection with high morbidity and mortality. Three most frequent and severe complications of IE are heart failure, paravalvular complication, and embolic events. This is a case of severe complicated IE having complete heart block (CHB) post-surgery.
Case Report: A 45 years old man, admitted to emergency unit with dyspnea, intermittent fever since months ago and signs of right heart failure. His past medical history included severe aortic regurgitation. Electrocardiogram revealed AF with some episodes of non-sustained VT. TTE showed dilation of all heart chambers, severe tricuspid regurgitation with multiple-mobile vegetation, severe aortic regurgitation with vegetation at aortic valve. Blood culture showed S.epidermidis, non positive result for major

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criteria. Diagnosis of definite IE was then established. Urgent heart surgery was then performed. Surgical findings revealed ruptured of the right sinus of Valsalva with fistula connecting to tricuspid annulus. Further, evacuation of vegetation, reconstruction of tricuspid and aortic annulus, and repairing ruptured sinus-of-Valsalva were done. Post surgery, the patient condition was relatively stable, however ECG revealed CHB. Pacemaker had been implanted to obtain adequate heart rate.
Discussion: Diagnosis of definite IE was based on the Modified Duke's Criteria. One major criteria was the echocardiography features of vegetation and fistula. Three minor criteria include AR as predisposing heart condition, fever, and positive blood culture not met major criteria. Complication of IE including fistula and large vegetation is the indication for urgent surgery as stated in 2016 ESC guidelines of IE management. TAVB was occurred due to structural deterioration affected AV node or wide surgical manipulation. Urgent surgery and immediate permanent pacemaker implantation improved early outcome.
Conclusion: IE remains a serious problems which can lead to severe complication and even mortality. Thus, appropriate treatment should be started immediately to have better clinical outcome.

P1060 - THE PROTECTIVE ROLE OF HYPOXIA INDUCIBLE FACTOR IN CHRONIC HYPOXIA INDUCED RIGHT VENTRICULAR REMODELING

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Objective: Generally, chronic hypoxia (CH) is a crucial factor in the development of congenital heart disease (CHD), especially in cyanotic congenital heart disease (CCHD). Clinically, right ventricular remodeling (RVR) is one of the most important determinants of longevity with certain CHD patients and CH-induced RVR correlates with an even worse aftermath. So improved understanding of molecular pathways such as hypoxia inducible factor (HIF) that modifies RVR may provide important clues toward novel therapies for CHD and RVR treatment.
Methods: Adult male Wistar rats (4wk) were placed in a chamber maintained at 10% O2 for 4 weeks. The chamber was constantly flushed with room air to maintain low (<0.5%) CO2 concentrations. A Real-time control system monitored O2 levels and injected 100% N2 as needed to maintain 10% ± 0.5% O2. Normoxic animals were kept in normal air (21% O2). Every single day of the hypoxic exposure, rats were weighed and injected daily with Dimethyloxalylglycine (DMOG) (100 mg/kg i.p.), which was diluted in five times the volume of sterile saline, or injected with an equal volume of sterile saline. In addition, we explored the response of myocardial erythropoietin and its receptor to hypoxic exposure.
Results: By comparison between the experimental group and the control group, treatment with DMOG attenuated myocardial fibrosis, apoptosis, and oxidative stress, which lead to enhanced RV contractile function. As an endpoint of HIF-dependent cardioprotection, a novel pathway in which nuclear factor kappa B links HIF-1 transcription was defined.
Conclusions: DMOG is a competitive inhibitor of hypoxia-inducible factor (HIF)-hydroxylated prolyl hydroxylase and has been shown to play an important role against ischemia-reperfusion myocardial injury. And this study supports a role for HIF-1 stabilizers in the treatment of RVR by improving the level of expression of VEGF, HO-1 and Glut-1. In addition, RVR follows a linear relationship with increased RV afterload.

P1063 - DYSLEXIA SENSORY INTEGRATION AND ACADEMIC PERFORMANCE IN MEXICAN CHILDREN WITH CONGENITAL HEART DISEASE

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The aim of this work is to find out if there is a relationship between Congenital heart disease(CHD), reading and writing difficulties (dyslexia), academic performance and sensory integration in children between the ages of 6 and 12 years old.
Materials and Methods: We had 30 participants : 15 children with Congenital Heart Disease, and 15 children with other pediatric diseases, but no chronic. The DST-J test for dyslexia detection in children, was administered to both groups and compared with their individual academic achievement. All the parents answered the Sensory Integration Questionnaire. None of the children had been diagnosed with a neurological or genetic disease. The DST-J test has been tested and validated with mexican children.
Results: For the data analysis we used the SPSS program and obtained a Pearson coefficient for each group and variables of the DST-J test related with the report card from the school in which each child is attending; specifically spanish, math and the general average of all the academic subjects. Concerning the Sensory Integration Questionnaire, 33.3% of the children with congenital heart disease have problems, compared with 13.3% of the other pediatric group. Regarding the incidence risk for dyslexia, the congenital heart disease group was 30% risk while the pediatric group 20%, with the same score for non risk children. The highest correlation between the predictive value for incidence of dyslexia and the subtests of the DST-J test was reverse digit, that was also highly correlated with the academic performance average marks. An intervention program was proposed considering the individual and group results. More studies need to be done comparing with normal population and bigger samples of children with complex congenital heart defects.

P1067 - CUSTODIOL SINGLE DOSE IS COMPARABLE TO BLOOD CARDIOPLEGIA FOR MYOCARDIAL PROTECTION IN CONGENITAL CARDIAC SURGERY

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Background: Custodiol used for myocardial protection due to single dose administration. We investigated single dose Custodiol myocardial protection versus intermittent blood cardioplegia in congenital cardiac surgery.
Methods: single-center retrospective review of prospectively collected data. One hundred twenty-seven patients <12 months undergoing biventricular congenital cardiac repair under cardiopulmonary bypass (CPB) were randomized to single dose Custodiol (n = 101) or blood cardioplegia (n = 26). Demographic, preoperative variables and postoperative (24 hours and discharge) echocardiography cardiac function were compared.
Results: same baseline patient's STAT mortality categories and score. Average CPB time (Custodiol: 90.7 ± 51.3 minutes vs blood cardioplegia: 82.6 ± 45.1 minutes, P = 0.46) and aortic cross-clamp time (Custodiol: 57.5 ± 33.1 minutes vs blood cardioplegia: 53.8 ± 34.3 minutes, P = 0.62) were similar. Patients on

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Custodiol were younger (101.3 ± 56.6 days), lower weight (4.8 ± 1.3 kg), compared with 143.6 ± 81.4 days and 5.4 ± 1.6 kg blood cardioplegia ($p < 0.05$). Left ventricle Ejection Fraction pre-operative (blood cardioplegia: 69.6 ± 14.6% vs. Custodiol: 73 ± 9.5%, $p = 0.15$), post-operative at 24 hrs (blood cardioplegia: 64.7 ± 10.6% vs. Custodiol: 65.1 ± 13.7%, $p = 0.89$), and post-operative at discharge (blood cardioplegia: 66.3 ± 8.5% vs. Custodiol: 67.7 ± 10.7% $p = 0.54$) were similar. The pre-operative right ventricle (RV) function by fractional area change (FAC) was lower in blood cardioplegia than Custodiol (blood cardioplegia: 43.4 ± 12.9% vs. Custodiol: 49.1 ± 8.2%, $p < 0.05$) although both normal (>40%).The FAC (24 hours and discharge) was 41.4 ± 12.7% and 41.9 ± 9.9% for blood cardioplegia, compared with 50.1 ± 9.1% and 50.9 ± 8.5% for Custodiol ($P < 0.001$).
Conclusions: Single dose Custodiol is convenient and as safe as blood cardioplegia for myocardial protection in congenital cardiac surgery with better RV function preservation.

P1072 - TRANSPOSITION OF GREAT ARTERIES WITH AORTOPULMONARY WINDOW (OPERATIVE VIDEO OF ARTERIAL SWITCH OPERATION)

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Background: Transposition of the great arteries with aorto-pulmonary window is a rare congenital cardiac anomaly. An arterial switch operation with repair of the aortopulmonary window is the preferred operation in this subset. As the tissue between the great arteries is missing, it is considered to be a complex operation. The purpose of the video is to demonstrate our simple yet highly effective surgical technique for arterial switch operation for this rare complex cardiac defect.
Methods: The video will demonstrate Arterial switch operation with minor modification in excision of branch pulmonary arteries is all that is needed in approaching this complex subset. The moiety of tissue resulting from the absence of an aortopulmonary window was naturally covered by the proximal and distal neoaortic flap tissue. The operative technique used in the 4 cases.
Results: Four patients of ages 28 days, 35 days, 40 days, and 6 months were successfully operated. One patient expired on postoperative day 21. In this case, advanced age of presentation, severe pulmonary artery hypertension, and sepsis possibly caused the death. The remaining 3 patients are off medication now and are being regularly followed up.
Conclusions: In our experience early diagnosis and an arterial switch operation have been crucial in getting a favorable outcome in planning of this complex congenital heart disease.

P1073 - A NEW SURGICAL TECHNIQUE FOR CLOSURE OF APICAL MUSCULAR VENTRICULAR SEPTAL DEFECT

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Objective: Ventricular septal defect (VSD) is among the most common congenital heart diseases encountered in pediatric cardiac patients. Apical muscular VSD constitutes nearly 2% of defects, which may or may not be associated with other congenital heart defects. The purpose is to present our innovative and simple surgical technique using custom-made low-profile polytetrafluoroethylene (PTFE) single disc device for closing multiple apical muscular and isolated apical muscular VSD.
Method: Between January 2010 and July 2016, 34 patients with isolated or multiple apical muscular VSDs with or without associated heart diseases underwent operation at our institute. The apical VSD was closed using our custom-made low-profile single disc polytetrafluoroethylene device. The operative technique and the technique used to prepare the single disc device will be presented.
Results: Thirty four patients of ages ranging from 3 months to 8 years underwent operation over 6 years. One 8-month-old patient with transposition of the great arteries with multiple VSDs died after 35 days due to severe pulmonary artery hypertension and sepsis. Another newborn infant with infracardiac total anomalous pulmonary venous connection with a 4-mm apical VSD also died after 3 days because this VSD could not be identified. All other patients are doing well on follow-up.
Conclusions: The technique described by us has the advantage of apical single/multiple VSD closure through the left ventricle without left ventriculotomy. Our technique is simple and cost-effective.

P1074 - THREE DIMENSIONAL PRINTING MODEL IN DOUBLE OUTLET RIGHT VENTRICLE UTILITY OF PATIENT SPECIFIC PATCH BAFFLE FOR INTRAVENTRICULAR REPAIR

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Background: Three-dimensional (3D) engineered replicas of different anatomical structures have been used extensively in different fields of medicine over the past years and are rapidly gaining interest in congenital heart surgery for surgical planning. We report a 6 year old patient with double outlet right ventricle, subtype Fallot, who underwent biventricular surgical repair with intracardiac baffle, right ventricle outflow tract augmentation and pulmonary valve comisurotomy after surgical planification with assistance of a 3D printed heart and patch model.
Material and Methods: Transthoracic echocardiogram, cardiac catheterization and computed tomography angiography (CTA) with 3D reconstruction of intracardiac anatomy were performed to plan surgical strategy for repair. Subsequently, 3D heart model and virtually created baffle patch were printed to asses repair suitability and strategies of baffle construction. Both the 3D printed heart and patch model were sterilized for intraoperative use to precisely cut the Dacron patch for baffle construction for intraventricular repair.
Results: The 3D heart model accurately depicted the ventricular septal defect position (VSD) relative to important adjacent structures, including the outflow tracts, aortic and tricuspid valves. Virtual planning, simulation and 3D printing of patch model were useful for Dacron patch trimming for baffle construction. Post-operative predischarge transthoracic echocardiogram revealed appropriate VSD baffling, without obstruction.

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Conclusion: Three-dimensional printed heart model of the intracardiac anatomy and patch simulation and printing using CTA data can provide valuable information for preoperative planning in DORV subtype Fallot repair. 3D printed patch templates are useful to simplify and increase accuracy for complex procedures.

P1075 - EMPOWERING THROUGH EDUCATION: SINGLE VENTRICLE CONSIDERATIONS FOR NON CARDIAC PROCEDURES

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Each year in the United States, 1 in 20,000 live births results in an infant born with complex single ventricle disease1. Surgical palliation of these defects usually involves three procedures. The first performed soon after birth, second at about six months and the third by three years. With advances in their management, many children are surviving beyond the third stage of palliation. Such an increase in life expectancy has resulted in an additional need for non-cardiac interventions including PICC lines, GI, ENT and radiologic procedures. The operating room nurse requires additional understanding of the physiologic and anatomic differences in patients with single ventricle physiology to maintain safety during non-cardiac surgical procedures. The objective is to empower the nurse through education to utilize their critical thinking skills. To achieve optimal safety, this education will aid in the integration of single ventricle considerations with high quality nursing standards. Increased knowledge and awareness will provide the nurse with confidence to administer individualized care to ensure a safe patient outcome.

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P1086 - SINGLE VENTRICLE PALLIATION AMONG DEVELOPING WORLD CONGENITAL HEART SURGERY PROGRAMS A REPORT FROM THE INTERNATIONAL QUALITY IMPROVEMENT COLLABORATIVE

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Objectives: Surgical care for congenital heart disease in the developing world is increasingly available. We describe patient characteristics, surgeries, outcomes, and risk factors for mortality among patients undergoing single ventricle palliation (SVP) procedures in this setting.
Methods: We identified SV diagnoses and Glenn or Fontan operations from a multicenter developing world database (32 programs in 20 countries) in 2010-2014 collected for quality improvement purposes. Clinical information and outcomes were summarized for each procedure. Multivariate models were used to identify risk factors for in-hospital mortality.
Results: Among 2,543 cases, there were 5.2% Stage 1 (S1), 5.9% pulmonary artery band (PAB), 7.2% systemic-to-pulmonary shunt (SPS), 26.5% Fontan, and 53.4% Glenn. Two-thirds had SV diagnoses: 19.8% tricuspid atresia, 16.5% double-inlet ventricle, 5.9% hypoplastic left heart syndrome, and 5.9% pulmonary atresia/intact ventricular septum. One-third had potential 2 V anatomy, including 14.0% double-outlet right ventricle and 9.1% transposition of the great arteries. SVP surgeries represented <1-15.6% of a program's congenital heart surgical volume. Overall survival was 89.3%, but mortality was considerable among initial palliations (S1 51.5%, SPS 24.6%, PAB 12.0%). In contrast to developed countries, a large proportion (61.3%) of initial palliations (S1, SPS, PAB) were performed after 1 month of age, and malnourishment and infections were common among them (47.0% and 19.8%, respectively). In addition to type of surgery, independent risk factors for death included age (≤ 30 days odds ratio (OR) 2.7, 31 days-1 year OR 1.9, $p < 0.005$), major medical illness (OR 2.2, $p = 0.003$), major non-cardiac structural anomaly (OR 2.1, $p = 0.009$), and low weight or body mass index for age (OR 1.8, $p < 0.001$).
Conclusions: We report outcomes from SV procedures performed in developing word settings, including strategies to perform Glenn or Fontan procedures in complex 2 V patients. Surgeries in neonates were rare, and malnutrition was common and increased the risk of death.

P1095 - UNUSUAL PRESENTATION OF AORTIC ARCH ANOMALY IN MONOZYGOTIC TWINS

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Anomalous origin of the innominate artery from the pulmonary artery is an extremely rare congenital heart disease that has been scarcely reported. Twin number 1, (11 month-old), presented with significant asymmetry in upper left side of the body, cervical hemivertebrae, scoliosis, and inspiratory and expiratory stridor. A two-dimensional ultrasonography and color doppler evidenced dilated left cavities without intra-cavitary pathology, right aortic arch and a continuous flow vessel communicating left subclavian artery with the pulmonary artery. Multislice CT angiography with 3D reconstruction demonstrated a right aortic arch and anomalous innominate artery emerging from the trunk of the pulmonary artery, and hypoplastic ipsilateral internal carotid artery resulting in an asymmetric intracranial flow. The angiographic procedure revealed significant circulation steal from the pulmonary artery trunk to the innominate artery resulting in a decrease on the left side cerebral blood flow. In twin number 2, an echocardiogram showed right aortic arch, severe stenosis at the origin of the left pulmonary artery with a gradient of 50 mmHg. CT angiography confirmed right aortic arch causing compression of the right main

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bronchus, causing an upper lobe chronic atelectasis, and left pulmonary artery with significant stenosis in the origin and abnormal length, with unapparent upper lobe branch. In twin number 1, resection of the left innominate artery with end-to-side anastomosis to the aortic arch and aortopexy was performed. In twin number 2, left pulmonary artery augmentation with pericardium patch and aortopexy resolved the chronic atelectasis. Both patients had good clinical outcome.

P1106 - ANALYSIS OF LONG TERM OUTCOMES OF CHILDREN CONGENITAL HEART DISEASE COMPLICATED WITH SEVERE PULMONARY ARTERIAL HYPERTENSION AFTER SURGERY

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Purpose: To analyze the survival status of children out of indication and to discuss the indication through follow-up on children with severe pulmonary hypertension associated with unrestrictive left-right shunt congenital heart disease.
Methods: Hemodynamic data and other information are collected from children with severe pulmonary arterial hypertension associated with congenital heart disease and treated in Anzhen hospital from January 2005 to June 2016. The survival analysis and log-rank test are executed as well as ROC curve.
Results: The object of observation is 59 children (aged 8.4 ± 5.2 years), 27 males and 32 females included, with major cardiac malformations of unrestrictive left-right shunt congenital heart disease, and follow-up time of 4-127 months (mean 69.3 ± 33.5 months). 12 children belong to Group PVRI $<$ 6 wood*U*m2 (6 male and 6 female), 3 lost to follow up, and none patient with postoperative PAH. Comparatively, 47 patients belong to Group PVRI $\geq$ 6wood*U*m2 (21 male and 26 female), 15 lost to follow up, and most of whom still suffered pulmonary arterial hypertension (one of them died 10 months after surgery) except 4 patients. According to postoperative living quality, the prognosis of Group PVRI $\geq$ 6wood*U*m2 with the rate of no positive event 86.6% in 3-year, 79.2% in 5-year and 22.5% in 10-year was significantly worse than group PVRI $<$ 6wood*U*m2 (Log-rank test $p = 0.0496$). Moreover, PVRI may further rise if surgeries are operated on the children out of indication. ROC curve demonstrates that PVRI is a sensitive indicator to predict post-operative pulmonary arterial hypertension(AUC 0.90), with the sensitivity of 84.2%, the specificity of 80.0%, as the boundary point of PVRI 8.15 wood*U*m2.
Conclusion: 1. long-term surgical outcomes is better in Group PVRI $<$ 6wood*U*m2 than in Group PVRI $\geq$ 6wood*U*m2. 2. PAH may progress in long-term post-operative follow-up if PVRI is too high before surgery. 3. Radical surgery is not recommended when PVRI is higher than 8.15 wood*U*m2.

P1109 - ACQUIRED VON WILLEBRAND SYNDROME IN INFANTS WITH AORTOPULMONARY SHUNT

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Background: The acquired von Willebrand syndrome (aVWS) was first described in 1968 by Simone and colleagues in patients with autoimmune diseases. The aVWS is very rare in children, most frequently being described in connection with congenital heart defects including aortic stenosis, PDA, VSD and pulmonary hypertnsion. The aVWS often results in increased bleeding tendency. Until now, there are no reports describing aVWS in infants with aortopulmonary shunts.
Patients and Results: Between 07/15-07/16 we evaluated 11 infants younger than 3 months with univentricular hearts and aorto-pulmonary shunt (9x Blalock-Taussig-Shunt, 1x central aorto-pulmonlary shunt, 1x Sano-Shunt (from the systemic right ventricle to the pulmonary artery)) and tested for avWD. The shunt operation was performed between day 5.-180. (median 8d), the blood samples were collected between days 18-260 after surgery (median 32d). In all 11 patients we identified avWD with a reduction/loss of the largest VWF multimers.
Discussion: Despite the limited number of patients, we can presume that nearly 100% of the patients with aortopulmonary shunt present aVWS. Its pathogenesis is explained by the increased activation of the VWF under the influence of the turbulent flow within the shunt. The activated VWF is bound to its specific receptors located on the platelets and on the activated endothelial cells, and undergoes an ADAMTS 13 mediated proteolysis, which leads to the loss of large multimeres. First results show that the VWF swiftly normalizes shortly after suppression of the shunt dependent lung perfusion and switching to a cavopulmonary (Glenn) connection.
Conclusions: So far none of our patients demonstrated an increased bleeding tendency in everyday life. However, we must consider this anomaly as a potential cause of increased blood loss during cardiac catheterizations and operations. Knowledge of the existence of an avWS is therefore necessary for introduction of the replacement therapy with FVIII/VWF products.

P1112 - MONOCUSP VALVE PLACEMENT DURING COMPLETE REPAIR OF TETRALOGY OF FALLOT WITH TRANSANNULAR PATCH DOES NOT HAVE BENEFITS AT 1 YEAR FOLLOW UP

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Background: A monocusp valve (MV) may be placed in the right ventricular outflow tract (RVOT) during complete repair of tetralogy of Fallout (TOF) using a transannular patch (TAP). There is limited data regarding outcomes of those with a MV and no data directly comparing those with and without a MV. This study compares 1-year follow-up data of children having undergone complete repair of TOF utilizing TAP with and without MV.
Materials and Methods: Children who underwent complete repair of TOF using TAP at the Children's Hospital of Wisconsin between the years 2000 and 2016 were identified. Clinical and surgical data was collected. Echocardiographic data from the pre-operative period, at time of discharge, and 12 months after repair was collected. Clinical, surgical, and echocardiographic data was then compared between those with and without monocusp.
Results: A total of 48 patients were included in this analysis, 30 (63%) with MV. There was no difference in preoperative characteristics. Peak RVOT gradient was greater in those with a MV (82.1 vs 67.0 mmHg, $p = 0.007$) although no difference was noted in preoperative PI. MV was surgeon dependent. Bypass time, cross clamp time, extubation in the operating room, and chest closure did not differ. Duration of mechanical ventilation, chest tube drainage, or postoperative hospital stay did not differ. Those without a MV were more likely to have moderate or severe PI at

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the time of discharge (83% vs 23%, p = 0.001) although there was no difference in peak RVOT gradient. At 1 year follow-up there was no longer a difference in PI and there was no difference in peak RVOT gradient. Need for reintervention and mortality at 1 year were also similar.
Conclusion: MV does not decrease PI, reintervention, or mortality at 1 year follow-up. A larger study with longer follow-up is warranted.

P1116 - COMPLETE REPAIR OF TETRALOGY OF FALLOT IN THE NEONATAL VERSUS NON NEONATAL PERIOD A META ANALYSIS

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Background: The surgical intervention of tetralogy of Fallot has evolved from a palliative intervention to a goal of complete repair. Optimal timing for complete repair, however, is not clear. For those who are asymptomatic, delayed complete repair in the first year of life is reasonable. For those who are symptomatic it is unclear if a staged palliation, including a neonatal shunt followed by complete repair outside of the neonatal period or complete repair within the neonatal period is preferable. We conducted a meta-analysis comparing neonatal and non-neonatal repair.
Materials and Methods: Several databases were queried to identify studies comparing neonatal and non-neonatal repair for tetralogy of Fallot. Manuscripts were reviewed for quality and bias with favorably scored manuscripts being included in the final analysis. A meta-analysis was conducted using the Mantel-Haenszel approach. Several perioperative and postoperative variables were compared.
Results: A total of 8 studies with 3,858 patients were included in the analysis. Of these patients, 19% underwent neonatal complete repair. Those who underwent neonatal complete repair had longer intensive care unit stays and longer total hospitalization. Neonatal repair was also associated with increased likelihood of need for transannular patch, delayed sternal closure, significant postoperative arrhythmias, and mortality.
Conclusion: Neonatal complete repair of tetralogy of Fallot is associated with increased morbidity and mortality. For those who are asymptomatic, complete repair should be deferred until outside of the neonatal period.

P1118 - TIMING OF FONTAN COMPLETION IN CHILDREN WITH FUNCTIONALLY UNIVENTRICULAR HEARTS AND ISOMERISM THE IMPACT OF AGE WEIGHT AND PRE FONTAN ARTERIAL OXYGEN SATURATION

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Background: Isomerism, or heterotaxy, impacts morbidity and mortality after various stages of univentricular palliation. Timing of Fontan completing in these patients based on preoperative factors has not been investigated previously. The aim of this study was to determine the impact of preoperative factors on various outcomes including length of hospital stay and duration of chest tubes.
Materials and Methods: A cross-sectional study was conducted. Patients with isomerism having undergone Fontan at the Children's Hospital of Wisconsin between 1998 and 2014 were identified. Preoperative, operative, and postoperative data was collected on these patients. Linear regression analysis was conducted to determine preoperative characteristics associated with various postoperative outcomes. Receiver operator curve analysis was also performed to determine the sensitivity and specificity of age and pre-Fontan arterial oxygen saturation in predicting increased length of hospitalization and increased duration of chest tubes.
Results: Younger age and lower pre-Fontan arterial oxygen saturation were associated with increased length of hospitalization while younger age, lower pre-Fontan arterial oxygen saturation, interrupted inferior caval vein, and worse pre-Fontan atrioventricular valve regurgitation were associated with increased length of chest tubes.
Conclusions: Age, arterial oxygen saturation, pulmonary arteriovenous malformations, interruption of the inferior caval vein, and atrioventricular valve regurgitation should all be taken into consideration when timing Fontan completion in patients with isomerism. Arterial oxygen saturations between 82 and 84% with an approximate age of 3 years appear to be a time at which it is reasonable to consider Fontan in patients with isomerism.

P1121 - ARTERIAL SWITCH SURGERY A 4 YEAR EXPERIENCE OF A SMALL CARDIAC CENTER IN BRAZIL

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Background/Hypothesis: The typical survival rate for infants with transposition of the great arteries (TGA) is greater than 90%. We aimed to evaluate the long term survival of all patients with TGA that underwent the arterial switch operation (ASO) at our small cardiac center in Brazil.
Material and Methods: Between April 2012 and October 2016, 28 consecutive patients who underwent ASO for TGA at Hospital Infantil e Maternidade Dr Alzir Bernardino Alves were included in this retrospective study. The median weight was 3.3 kg, median age 14.5 days, 21 were male and 7 female. Following parameters were evaluated: use or non-use of custodiol for cardioplegy, size of the post operative care team (small, 4 physicians or large, 8 physicians), simple TGA (TGA with intact ventricular septum) and complex TGA (TGA with ventricular septal defect or others anomalies) For statistics analisys were used the Kruskal-Wallis test, the Cox hazard proportion and Kaplan Meier curve.
Results: Overall mortality was 28.5% (8 patients), however, in the period after July 2014 we operated 18 patients with only one death (5.5%). The comparison between small team vs large team and between with versus without custodiol was statistically significant ($p < 0.05$), showing the improvement on mortality rate with the large team and with custodiol. Accordingly with Cox hazard proportion the non-admission of the custodiol represented a major risk factor to mortality ($p < 0.05$). Associated heart disease and coronary anomaly showed no significant difference for survival.

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Analyzing the patient's death during the period, 5 occurred on the first 24 postoperative hours (PO), 2 in the first 5 days and 1 in the 14th PO. We have 20 live patients in clinical follow-up.
Conclusions: The observed survival rate was 71.5%, with significant improvement after the first 2 years. The increase of post operative team and the use of custodiol were the key to achieve the observed survival improvement.

P1122 - DETERIORATION OF FUNCTIONAL ABILITIES AND STROKE IN CHILDREN SURVIVING THE FONTAN PROCEDURE

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Background/Hypothesis: Children surviving complex cardiac surgery are at risk for developmental delays. We hypothesized deterioration of functional abilities (DFA) is not uncommon after the Fontan procedure, and often due to stroke. Objectives: To determine the frequency of 1) DFA post-Fontan 2) post-Fontan stroke among those with and without DFA.
Materials/Methods: As part of The Western Canadian Complex Pediatric Therapies Follow-up Program (CPTFP), from 1996-2016, 187 children prospectively registered in the CPTFP underwent a Fontan operation at Stollery Children's Hospital. At age 2 and 4.5 years, surviving children received multidisciplinary assessment, and the Adaptive Behavior Assessment System-II general adaptive composite (GAC) was determined. DFA was defined as a 1 SD decrease in GAC in comparison to pre-Fontan scores. Stroke diagnosis was ascertained through retrospective chart review. Frequency of DFA and stroke are presented as percentage of assessed survivors. T-tests and chi-square tests compared groups.
Results: Results are obtained for 156/181 survivors (64% male, 52% classic hypoplastic left heart syndrome). DFA occurred in 38/156 (24.4%), and stroke occurred in 16/156(10.3%). Mean (SD) post-Fontan GAC in all children with stroke was 77.4 (22.3) vs. 89.6(18.7) in those without stroke ($p = 0.01$). Evidence of post-Fontan stroke was found in 9/38 (23.7%) of children with DFA, vs. 7/118 (6%) of children without DFA ($p = 0.002$). Mean (SD) GAC decline among children with DFA was greater in those with stroke [25 (9.6)] than in those without [20 (6)], though the difference was not statistically significant ($p = 0.16$).
Conclusion: Almost 25% of children surviving the Fontan procedure show DFA, which has major implications for their functioning later in life. Evidence of post-Fontan stroke is more commonly found among those with DFA; however, stroke is not the only event leading to DFA. The next step is to determine potentially modifiable predictors of DFA.

P1128 - IS THE PRESENCE OF END DIASTOLIC FORWARD FLOW SPECIFIC FOR A RESTRICTIVE RIGHT VENTRICULAR PHYSIOLOGY IN A REPAIRED TETRALOGY OF FALLOT

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Background: The presence of end-diastolic forward flow (EDFF) has been regarded as a r-RVP, but conflicting results have been reported regarding the effects of r-RVP on the clinical outcome in patients with r-TOF.
Objectives: We hypothesized that the EDFF by Doppler was not specific for the diagnosis of r-RVP.
Methods: Sixty-two consecutive patients aged 15.7 ± 11.6 years who underwent cardiac catheterization were studied. The patients were divided according to the presence of EDFF (group 1: EDFF + , group 2:EDFF-) and right ventricular (RV) size (group A: small RV, group B: large RV [> 150 ml/m2]). We compared clinical and hemodynamic variables between the groups.
Results: The EDFF with at least 3 consecutive and >30 cm/sec velocity was present in 23 of 62 patients (39.6%). Group 1($n = 23$) had higher right atrial pressure (RAP), pressure gradients between the RAP and pulmonary diastolic pressure (PDP), and atrial natriuretic peptide (ANP) levels than group 2. Four patients (17.4%) in group 1 and 89.7% in group 2 had a normal RAP range (a wave < 10 mmHg). There were no differences in the RV volume and ejection fraction (EF), left ventricular (LV) volume, LVEF, B-type natriuretic peptide (BNP) levels, cardiothoracic ratio, QRS duration and severity of pulmonary regurgitation (PR) between groups 1 and 2. Group A had a better RVEF, LVEF and smaller LV size than group B. The RAP in subgroup 1 A was higher than that in the other 3 subgroups. Subgroup 1B had a similar RAP as group 2, and had lower PDP and more severe PR than the subgroup 1 A.
Conclusions. Patients with the presence of an EDFF are associated with an increased ANP, but not BNP. The presence of EDFF may not be specific for r-RVP. A more accurate definition is necessary to determine how the r-RVP effects the clinical outcome in r-TOF.

P1137 - COMPARISON OF SEPTAL LEAFLET DETACHMENT VERSUS CHORDAL DETACHMENT TECHNIQUES IN CLOSURE OF HARD TO EXPOSE VENTRICULAR SEPTAL DEFECTS

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Purpose: Different techniques have been used for exposure of ventricular septal defect (VSD) margins when there is crowding of the anatomy by tricuspid valve (TV) subvalvar apparatus. The aim of this study was to compare surgical outcomes, for the two techniques of TV leaflet detachment (LD) and the rarely described TV chordal detachment (CD) for hard-to-expose VSDs.
Methods: All patients undergoing transatrial VSD repair at our institution were identified from our database. Follow-up echocardiography and patient data were obtained from medical records. Between 1/2005-12/2015, 121 isolated perimembranous VSDs were repaired. Among these, 24 patients had LD, while 13 underwent CD, and 84 had regular VSD repair (reference group).
Results: There was no significant difference between the groups in age, weight, postoperative length-of-stay (LOS), inotropic score, genetic/syndromic abnormalities, pre- or postoperative gastric surgery, time-to-extubation, and discharge medication. The CPB and cross-clamp times were significantly higher in LD group, when compared with reference group (Table). Return to second CPB run occurred in 5 reference group patients, 1 LD, and no CD

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patient; these differences were not significant. Two patients (reference group) required pacemaker insertion. No operative deaths occurred. Follow-up echocardiograms were available for 76 patients at a mean of 2.6 years (1 month-11 years). TV regurgitation was rated as none or trivial in 59 (78%) and mild in 17 (22%). There was no difference in presence of residual VSD, or degree of TV regurgitation amongst the three groups. There was no reoperation for TV regurgitation. Two patients (reference group) underwent reoperation due to residual VSD, 1 and 3 years after initial surgery.
Conclusions: Tricuspid valve LD and CD techniques provide a viable and safe alternative to closure of hard-to-expose VSDs while maintaining appropriate TV function. Their use in our series did not lead to increased TV dysfunction at midterm echocardiographic assessment.

Table 1. Comparison between septal and chordal detachment and the reference group.

Variables	CD group (N = 13)	LD group (N = 24)	Reference group (N = 84)	CD/LD (P value)	LD/ Reference (P value)	CD/ Reference (P value)
Age (median, IQR, d)	174 (117-575)	187 (128-590)	140 (87-199)	0.23	0.34	0.70
Weight (median, IQR, kg)	6 (4.9-9.6)	6 (4.4-11)	5 (3.9-6.7)	0.23	0.24	0.96
CPB (mean +/-SD, min)	108+/-23	115+/-29	102+/-33	0.42	0.04	0.30
CCT (mean +/-SD, min)	67+/-17	70+/-20	60+/-24	0.46	0.03	0.22
LOS (median, IQR, d)	5 (4-7)	5 (4-11)	5 (4-7.5)	0.81	0.71	0.95

CCT = cross clamp time, CD = chordal detachment, CPB = cardiopulmonary bypass, LD = leaflet detachment, LOS = postoperative length-of-stay.

P1139 - PREDICTING FEASIBILITY OF BIVENTRICULAR REPAIR IN UNBALANCED ATRIOVENTRICULAR SEPTAL DEFECT

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Background: Unbalanced forms of atrioventricular septal defect continue to be challenging and present poor surgical outcomes. Echocardiographic indicators such as atrioventricular valve index, right ventricle/left ventricle inflow angle and size of the ventricular septal defect have been identified as relevant discriminators that may guide surgical strategy.
Methods: A new variable we called "indexed ventricular septal defect" is described, which reflects the size of the defect in relation to the atrioventricular valve diameter. We outline a geometrical description of the echocardiographic features of atrioventricular septal defect and derive an equation relating the new index with the atrioventricular valve index and the right ventricle/left ventricle inflow angle. In the light of that equation we discuss the interdependence between variables and employ data of the inflow angle presented in a study held by the Congenital Heart Surgeons' Society to set limits on the new index.
Results: For moderately unbalanced forms (modified atrioventricular valve index between 0.2 and 0.39), indexed ventricular septal defect can guide surgical decision-making: a) under 0.2, biventricular repair is strongly recommended; b) between 0.2 and 0.3-0.35, biventricular repair can be achieved depending on other factors; c) between 0.3-0.35 and 0.5-0.55, other strategies must be considered; and d) above 0.5-0.55, univentricular palliation must be followed.
Conclusions: Combined use of indexed ventricular septal defect and atrioventricular valve index can significantly narrow the grey zone of unbalance and guide surgical approach.

Table. Indexed VSD as a function of mAVVI and RV/LV inflow angle.

mAVVI/α	80°	90°	100°	110°	120°	130°	140°	150°	160°	170°
0.02	0.25	0.14	0.07	0.04	0.03	0.02	0.01	0.01	0	0
0.05	0.32	0.21	0.14	0.10	0.07	0.05	0.03	0.02	0.01	0
0.08	0.37	0.27	0.19	0.14	0.10	0.08	0.05	0.04	0.02	0.01
0.11	0.41	0.31	0.23	0.18	0.13	0.10	0.07	0.05	0.03	0.01
0.14	0.44	0.34	0.26	0.20	0.16	0.12	0.09	0.06	0.04	0.02
0.17	0.47	0.37	0.29	0.23	0.18	0.14	0.10	0.07	0.05	0.02
0.20	0.49	0.40	0.32	0.25	0.20	0.16	0.12	0.08	0.05	0.02
0.22	0.51	0.41	0.33	0.27	0.21	0.17	0.13	0.09	0.06	0.03
0.24	0.52	0.42	0.34	0.28	0.22	0.18	0.13	0.10	0.06	0.03
0.26	0.53	0.43	0.35	0.29	0.23	0.18	0.14	0.10	0.06	0.03
0.28	0.54	0.45	0.36	0.30	0.24	0.19	0.15	0.11	0.07	0.03
0.30	0.55	0.45	0.37	0.31	0.25	0.20	0.15	0.11	0.07	0.03
0.32	0.56	0.46	0.38	0.31	0.25	0.20	0.16	0.11	0.07	0.03
0.34	0.57	0.47	0.39	0.32	0.26	0.21	0.16	0.12	0.08	0.04
0.36	0.57	0.48	0.40	0.33	0.27	0.21	0.17	0.12	0.08	0.04
0.38	0.58	0.48	0.40	0.33	0.27	0.22	0.17	0.12	0.08	0.04

Different combinations of values of mAVVI and RV/LV inflow angle (α) are entered to obtain the respective inVSD in the uAVSD group. This number represents the size of the VSD indexed to the diameter of the AVV.

P1142 - PSEUDO ANEURYSM ERODING INTO THE CHEST WALL FOLLOWING AORTO PULMONARY SHUNT

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Background: Pseudo-aneurysms of ascending aorta,following surgery for congenital heart defects are exceedingly rare. Their usual location is at the aortic cannulation site. We present a first-ever case of pseudo-aneurysm following a central aorta-pulmonary shunt.
Material & Methods: An 11 year-old boy presented with acute onset pain and swelling near the sternal head of his right clavicle. Past medical history included a Tetralogy of Fallot (TOF) diagnosed in infancy, with central aorto-pulmonary shunt placement at age 6 years performed at another hospital. Computed Tomography delineated a large pseudo-aneurysm measuring 12X10 cm, arising from the ascending aorta and protruding under the upper end of sternotomy scar after eroding into the first rib, medial clavicular head and sternal manubrium. Emergency surgical correction was scheduled with a plan of concomitant repair of TOF.
Results: Cardiopulmonary bypass(CPB) was instituted using the right femoral artery and vein and cooling to 16 degrees C to achieve total circulatory arrest (TCA). Re-sternotomy was performed and the wall of pseudo- aneurysm was breached. The aorta-pulmonary shunt graft was found floating within the clots and altered blood. The defect in the ascending aorta measured 8X5 cm, and was repaired with a Polytetra fluroethylene (PTFE) patch. CPB was resumed after transferring the cannulae centrally, followed by complete intracardiac repair for TOF. The postoperative course was uneventful, requiring 12 hours of mechanical ventilation and hospital stay of 6 days.

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Conclusions: This is an extremely rare and previously unreported complication of a central aorta-pulmonary shunt. The aneurysm was contained by the surrounding structures initially, but increased in size gradually to erode into the chest wall. CPB using femoral cannulation, deep hypothermia and TCA is recommended to avoid fatal exsanguination during re-sternotomy for a large pseudo-aneurysm of ascending aorta.

P1167 - A NOVEL SYNTHETIC HYBRID CARDIOVASCULAR PATCH PROMOTES VIABLE TISSUE REGENERATION IN THE LARGE VESSELS

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Background/Hypothesis: Vascular wall regeneration has been attempted using various types of biodegradable materials containing stem cells, but the results have been disappointing. The aim of this study was to investigate whether vascular wall regeneration could be promoted by a unique synthetic cardiovascular patch (OFT patch).
Materials & Methods: The OFT patch is made with an extensible warp-knitted fabric composed of a combination of biodegradable (poly-L-lactic acid: PLLA) and non-biodegradable (polyethylene terephthalate: PET) yarn. The fabric is coated with gelatin to seal its porosity. The in vitro mechanical properties of the OFT patch were compared with those of commercial polytetrafluoroethylene (PTFE) and bovine pericardial patches. Part of the vascular wall of either the canine inferior vena cava (IVC) or descending aorta was replaced with an OFT patch. After 1, 3, 6, and 12 months, the patch was removed for histological examination.
Results: The OFT patch had excellent tensile strength, suture retention strength, and bending resistance that were equivalent to those of the two commercial patches. Furthermore, needle hole leakage from the OFT patch was far less than that from the PTFE patch. Microscopic examination revealed optimal tissue regeneration characterized by layered smooth muscle cells and collagen fibers induced in both the IVC and aorta after replacement. Interestingly, tissue connections developed across the OFT patch with neo-arterioles, along with disappearance of gelatin. There was a minimal inflammatory reaction, no calcium deposition on the patch yarn, and no aneurysmal or stenotic changes in patch-implanted regions.
Conclusions: The OFT patch had favorable mechanical properties and resulted in optimal tissue regeneration in the early stages after implantation in large vessels. Since the non-biodegradable portion is program-designed to expand to quadruple its initial size, the OFT patch would not interfere with vascular wall ingrowth as the biodegradable PLLA portion is absorbed.

P1171 - STAGED COMPLETE REPAIR WITHOUT HOMOGRAFT USE IN PATIENTS WITH PULMONARY ATRESIA–VENTRICULAR SEPTAL DEFECT AND MAJOR AORTOPULMONARY COLLATERAL ARTERIES

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Objective: Repairing pulmonary atresia–ventricular septal defect (PAVSD) and major aortopulmonary collateral arteries (MAPCAs) remains challenging, particularly without the use of a homograft. Here, we evaluated our surgical strategy, which consists of primary unifocalization with an autologous pericardial patch and staged complete repair using a handmade expanded polytetrafluoroethylene (ePTFE) tri-leaflet valve conduit.
Methods: Since 2007, eleven consecutive patients with PAVSD and MAPCAs underwent primary unifocalization. The median patient age and body weight were 7.3 months (2–116 months) and 7.3 kg (2.7–16.2 kg), respectively. All patients underwent primary unifocalization using tissue-to-tissue anastomosis. The central pulmonary artery was absent or diminutive in all cases; therefore, it was created using an autologous pericardial patch with a target diameter of at least 10 mm. The pulmonary blood flow source was established using a modified Blalock-Taussig shunt in 9 cases, a central shunt in 1 case and a right ventricle-pulmonary artery shunt in 1 case. The mean number of MAPCAs was 3.8.
Results: There was 1 operative death due to congestive heart failure after unifocalization. Post-unifocalization cardiac catheterization showed that the mean pulmonary artery (PA) resistance was 2.4 wood units and the mean PA pressure was 16.1 mmHg (13–24) under mean Qp/Qs = 1.05. Nine patients underwent staged complete repair within an average of 7.1 months after unifocalization. One patient is awaiting complete repair. The median body weight was 9.5 kg (6.8-17.6 kg). The median ePTFE conduit diameter was 16 mm (16–20 mm). The mean intraoperative right ventricle/systemic artery pressure ratio was 0.53 (0.4–0.71). Post-repair cardiac catheterization showed that the mean right/left ventricular pressure ratio was 0.58 (0.4–0.97).
Conclusions: Primary unifocalization was performed completely, even without a homograft. Acceptable right ventricular pressure was obtained. A sufficiently large handmade valve conduit to avoid early reoperation could be applied during the planned staged complete repair.

P1179 - PROPOSING GUIDELINES AND EVALUATION OF TWO STAGE ARTERIAL SWITCH AS A TREATMENT STRATEGY IN THE MANAGEMENT OF DELAYED PRESENTATION OF TRANSPOSITION OF THE GREAT ARTERIES WITH A REGRESSED LEFT VENTRICLE

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Background and Objectives: Management of transposition of the great arteries with intact ventricular septum (TGA/IVS) is currently an arterial switch operation (ASO) performed in the first 2 weeks of life. Two stage ASO is one form of treatment in infants with TGA presenting late. But the guideline on how to train LV is not clear in literature.
Methods: From December 2009 to January 2016 total of 33 patients with TGAIVS presented late and were selected for a two stage ASO. Serial echocardiography was used to assess the increased thickness of LV posterior wall. A stage II ASO was done a few weeks later. Retrospective review of patient charts was done. Data was formulated into a structured database and statistical analyses were performed with the statistical package SPSS for Windows.
Results: 33 patients underwent Stage I. 10 patients underwent isolated PAB, 17 patients underwent PAB and BTS and in 6 patients PDA stenting was done. Stage I had an in hospital

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mortality of 22%, while the interval mortality between both stages was 14%. Initial increased mortality was probably due to acute volume loading of the RV due to unrestrictive atrial communication, hence we modify to do PA band only in the presence of large ASD, and to shunt only selected patients. The mean interval between the two stages was 3 ± 1 weeks.16 patients have undergone a successful Stage II ASO. Stage II mortality was zero. All patients had remarkably rapid recoveries and short hospital (6 ± 2 days) stay.
Conclusion: Early experience indicates that in a developing country like Pakistan with a rapid two stage arterial switch is an acceptable treatment option. Patients who survived Stage I and the interval period have excellent results with Stage II.

P1180 - DIFFICULTIES AND POSIBLE SOLUTIONS IN ESTABLISHING PEDIATRIC CARDIAC CARE IN A DEVELOPING COUNTRY

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Background: Establishment of pediatric cardiac services in developing countries is a major challenge. Once established, it is even bigger a challenge to cater for the needs of very large populations. In Pakistan with population of 190 millions, about 4000 babies require urgent open heart surgery each year.
Objectives: To analyze difficulties and propose solutions, in establishing a pediatric cardiac program in a developing country.
Methods: The Children's Hospital Lahore is the only public sector children's hospital in the country where we established a congenital cardiac program and neonatal interventions and open-heart surgeries are being performed. It is a multidisciplinary hospital, with all the support facilities. In a developing country, there are numerous challenges like paucity of centers and personnel, limited resources, overcrowding, illiteracy, and infection control.
Results: Pediatric cardiac program at CH started in 1999. The problems faced include recruitment of trained staff and equipment and lack of resources for ongoing financial needs. So far we have performed surgery on >5000 children, with an overall mortality of 7% in the year 2015. The cost for an open heart surgery is about US $1200 and for a closed heart surgery is US $250. This is still too much for a poor patient. The current waiting list for routine open heart surgery is over 3 years.
Conclusions: Despite improved infrastructure and skills, the logistics, affordability, staffing, late presentation, nutritional status, infection, and unfavorable economics continue to negatively influence the overall results. With continued determination and philanthropic support, establishment of pediatric cardiac program is still possible in a developing country.

P1197 - MANAGEMENT OF PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM FOR OLDER PATIENTS A SINGLE CENTER EXPERIENCE FROM CHINA

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Background: Despite surgical treatment of the pulmonary atresia with intact ventricular septum (PAIVS) has been improved, the clinical outcomes of PAIVS patients with older ages were still deficient with related studies. In the present study, we summarized the experiences and evaluated the clinical outcomes of PAIVS patients with ages over 6 months for the first surgery in our heart center.
Methods: Between January 2006 to June 2016, 45 cases of PAIVS with ages over 6 months finished definitive repairs were reviewed. The patients were received definitive repairs, including biventricular repair, 1.5 ventricular repair and univentricular palliation without or with initial intervention. Median follow-up time was 5.3 years (range, 0.5 to 10.6 years), and survival, risk factors for death, and clinical status after definitive repairs were assessed.
Results: A total of 3 patients died (6.7%, 3/45) during follow up. Fontan procedure made up a large proportion of the older PAIVS patients (42.2%, 19/45), while only a few patients received biventricular repair (22.2%, 10/45) since the growth potential of RV was limited in the older patients. The medium-term (10 years) survival rates of biventricular, 1.5 ventricular and univentricular palliation were 100.0%, 93.3%, and 81.2%, respectively ($P > 0.05$). Moreover, multivariate analysis indicated that there were no independent predictors had the statistical difference as risk factors for death because of the low mortality in the present study. At latest follow-up, most patients had well clinical status after definitive repairs, while re-operation rate was 10.0% (1/10) after biventricular repair, 14.3% (2/14) after 1.5 ventricular repair and 20.0% (3/15) after univentricular palliation.
Conclusion: Favorable clinical outcomes of older PAIVS patients that managed with different definitive repairs were found in our retrospective study. In addition, the choice of optimal definitive repairs for the older PAIVS patients could also be achieved with a low mortality.

P1199 - SURGICAL OUTCOME OF CONGENITAL HEART DEFECTS WITH SEVERE PULMONARY HYPERTENSION IN INFANTS

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Background: Congenital heart diseases with large left-to-right shunt often complicated with severe pulmonary hypertension. It is an important determinant of morbidity and mortality in patients without educate surgical treatment especially in infants.
Methods: one hundred forty five patients with congenital cardiac septal defects and pulmonary arterial hypertension operated to close their septal defects. All the patients checked by Chest X-ray, EchoCG, ECG, selectively performed the cardiac catheterization and lung biopsy. Before and after surgery the PA pressure was compared to systemic by needle puncture measurement.
Results: Thirty-four patients died in the hospital after operation. 6 of them were under 6 months and others above 1 year. There were no later deaths in follow-up. Hemodynamic changes after operation included a significant decrease in pulmonary artery pressure (mean pulmonary artery pressure, 28.3 ± 2.4 mm Hg versus 58.45 ± 1.69 mm Hg before repair). The follow-up period was from 3 months to 4 years (mean 1.3 ± 0.6 years).
Conclusion: Studies from developed countries have shown that in term infants, young age is not a risk factor for adverse postoperative outcome after surgical closure of septal left-to-right defects. The main reason of the positive result is educate time for surgery and the level of cardiology service diagnostic developing. The data presented in this study show that operations to close cardiac septal defects in the presence of severe pulmonary hypertension are effective, but must done during first 6 monthes.

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P1206 - CASE REPORT PRIMARY REPAIR TETRALOGY OF FALLOT AND MAJOR AORTO PULMONARY COLLATERAL ARTERIES WITH SUSPECTED NOONAN SYNDROME IN DR. SOETOMO TEACHING HOSPITAL SURABAYA INDONESIA

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Background: Tetralogy of Fallot (TOF) is a complex congenital heart disesase including four abnormalities; ventricular septal defect (VSD), pulmonary stenosis (PS), right ventricular hyperthrophy (RVH), and overiding aorta. Cyanotic heart defect can develop Aorto-pulmonary collateral (APCA) to supply underperfused pulmonary bed. Genetic syndromes can occur in 20% of patients with TOF. Noonan syndrome is a common genetic disorder characterized by facial anomalies, congenital heart defect, short stature, webbed neck, chest deformities and cryptochidism. In developing country, diagnosed genetic syndromes associated with heart defect hasn't been covered by health insurance, yet the outcome of the primary repair still unknown.
Objective: To report the outcome primary repair tetralogy of fallot and major aorto-pulmonary collateral arteries with suspected Noonan Syndrome.
Case Report: A 11 years old boy admitted with cyanosis since birth, dispnea on effort, growth delay, learning problems, scoliosis, micropenis, hypertelorism, exopthalmos, facial dysmorphism, cryptochidism, and major aorto-pulmonaries collaterals. The patient undergone an echocardiographic study and it shows VSD malalignment with overiding aorta 48%, critical pulmonary stenosis, size of pulmonary arteries is favorable for primary repair. Cardiac

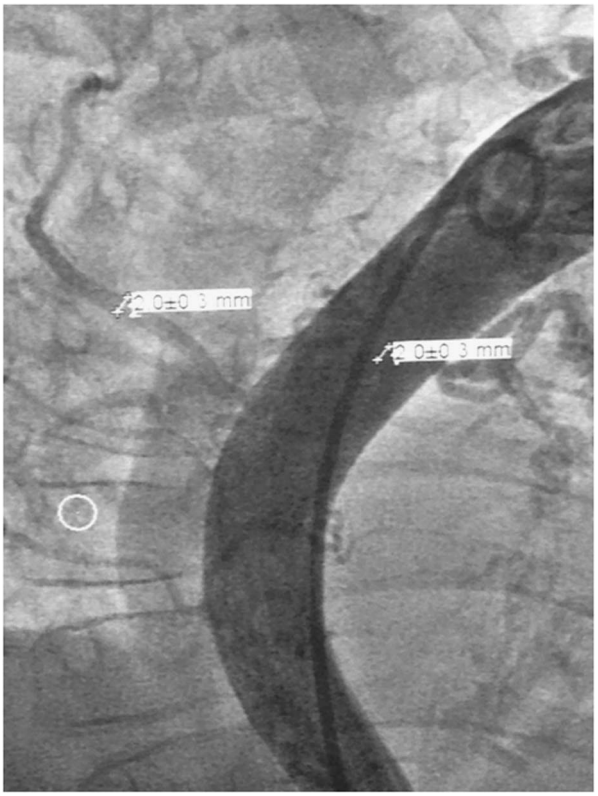

Figure 2.

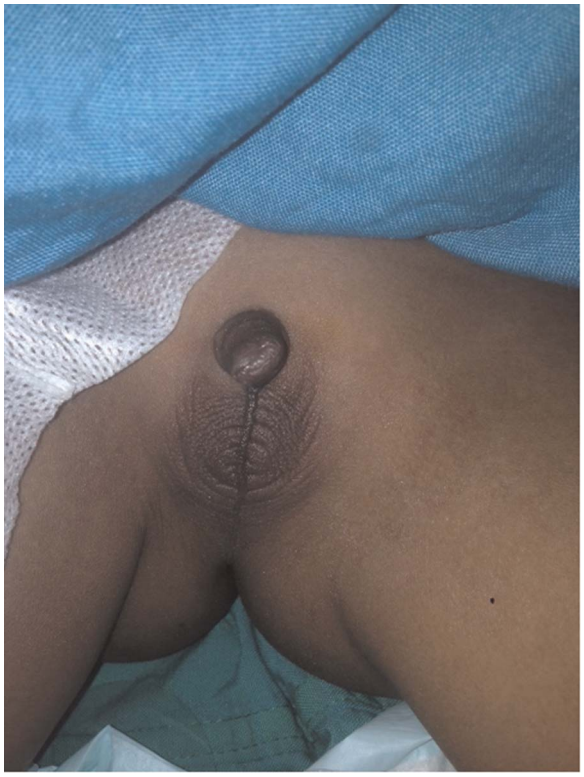

Figure 1.

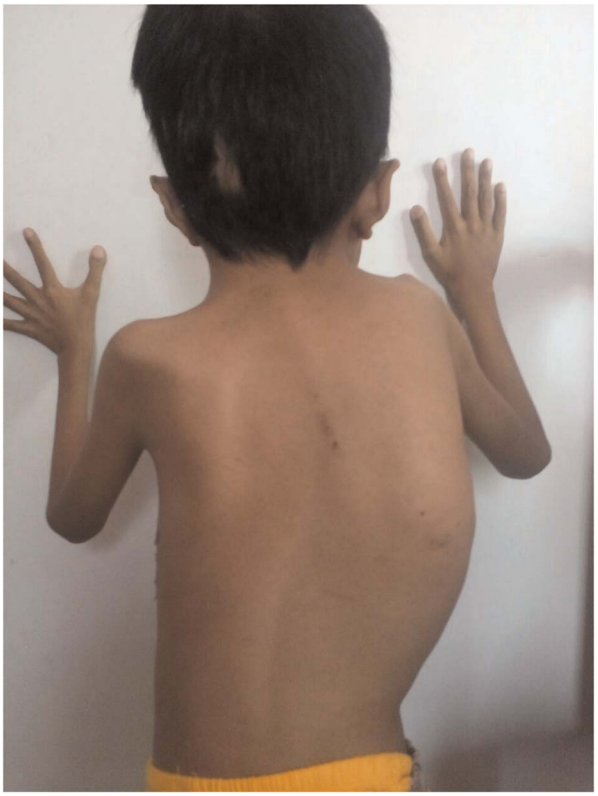

Figure 3.

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catheterization shows classic fallot with normal coronary arteries and major aortopulmonary collateral arteries (MAPCA) arising.
Surgical Technique: The patient underwent percutaneous coil embolization just before surgery and then median sternotomy was done and aortic cross-clamping are achieved, right atriotomy was performed and there provided exposure to malalignment VSD. A longitudinal pulmonary arteriotomy is made to expose infundibular stenosis. Infundibulectomy was performed and reconstruct MPA with pericardial patch. The VSD is closed using PTFE patch. The patient transferred to ICU afterward.
Result: Post operative echocardiography shows good outcome, and the patient was discharged after 15 days.
Conclusion: Primary Repair TOF with MAPCAs and suspected Noonan Syndrome was done successfully.

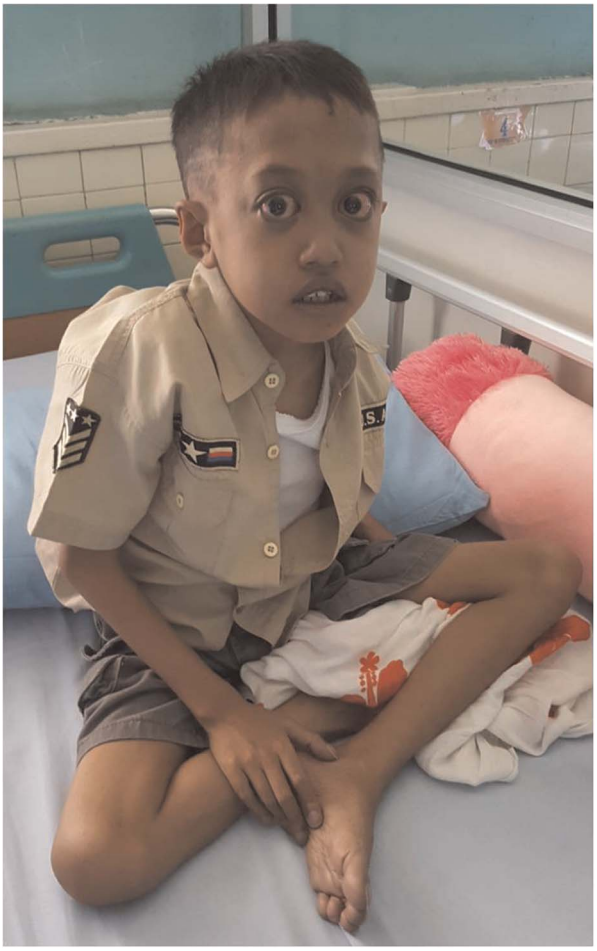

Figure 4.

P1220 - OUTCOMES OF HEPATIC VENOUS REDIRECTION FOR PULMONARY ARTERIOVENOUS MALFORMATIONS FOLLOWING KAWASHIMA PROCEDURE IN LEFT ATRIAL ISOMERISM

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Background: The development of pulmonary arteriovenous malformation (PAVM) after Kawashima procedure (TCPS) in left atrial isomerism (LAI) frequently induces desaturation, and then the redirection of hepatic vein to the pulmonary circulation (TCPC) improves oxygen saturation (SO2) with PAVM's regression. However, the degree of the improvement has not been unclearly delineated, and in addition, the effective timing of the conversion to TCPC remains unclear. The aim of this study is to examine the changes in SO2 and the occurrence of PAVM after TCPS and to propose the appropriate timing of the TCPC conversion.
Methods: Twenty-two patients with LAI achieving TCPC were reviewed. Antegrade pulmonary flow was completely interrupted during surgery of TCPS. Mean age at TCPS and TCPC were 6.6 ± 3.7 months-old and 1.5 ± 0.3 years-old, respectively. The ages at TCPS and TCPC of the patients with LAI are not different from those of patients with other types of univentricular physiology in our institute. PAVM was diagnosed by contrast echocardiography, catheterization or pulmonary angiographic findings as follows; 1) contrast agent return into the left atrium within 2-4 beats, 2) spongy pattern of pulmonary vasculartur, and 3) less than 92% of SO2 in pulmonary venous blood. Data of SO2 were collected at discharge from hospital, visit to outpatient clinic after TCPS, at catheter examinations before TCPC, and 1 year later after TCPC.
Results: SO2 decreased from $88.5 \pm 4.5\%$ to $79.1 \pm 10.1\%$ during the interval of 8.4 ± 4.2 months after TCPS and conversely increased to $92.3 \pm 3.7\%$ following conversion to TCPC ($p < 0.01$). The occurrence rate of PAVM was 77.3% about 8 months after TCPS and successfully decreased to 38.1% one year after TCPC ($p < 0.01$).
Conclusions: PAVM occurred with significant reduction SO2 even in the short interval of 8 months after TCPS. To prevent severe hypoxia and maintain oxygen level appropriately, therefore, TCPC would be performed within 1 year after TCPS.

P1221 - MID TERM SURGICAL OUTCOMES OF PARTIAL ATRIOVENTRICULAR SEPTAL DEFECT

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Background: Relatively high reoperation rate (17 to 25%) has been reported after repair of partial atrioventricular septal defect (pAVSD) mainly due to left AV valve regurgitation (LAVVR). We have developed a simple method with special focus on complete closure of the cleft. The purpose of the present study was to assess the outcomes of repair of pAVSD using this technique.
Methods: We reviewed 61 patients with including 5 with intermediate AVSD. Age and body weight were 3.8 ± 3.8 years old and 13.3 ± 9.9 kg, respectively. There were 16 patients with Down syndrome and 9 with polysplenia. At surgery wide coaptation zone was obtained with complete cleft closure up to the chordal attachment. Associated cardiovascular lesions included patent ductus arteriosus (4), coarctation of the aorta (2), ventricular septal defect (2), partial anomalous of pulmonary venous drainage (2), Cor triatriatum (1), and Lutembacher syndrome (1). Pacemaker (PM) was implanted in 2 patients with polysplenia before pAVSD repair. Median follow- up was 6.7 years (0.4-12.7 yeas).
Results: Median stays in ICU and hospital were 1 days (1~22 days) and 5 days (3~68 days), respectively. There were no hospital and three late deaths with non-cardiac causes in patients with polysplenia. Survival rate was $96.0 \pm 2.8\%$ at 5 years. Freedom from reoperation was $96.0 \pm 2.8\%$ at 5 years. Three patients (4.9%), who had complete repair at the age of less than 60 days old, underwent

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reoperation for subaortic stenosis, residual atrial septal defect, and LAVVR. The other two patients with polysplenia had PM implantation for SSS after repair.
Conclusions: Mid-term outcomes of complete repair of pAVSD using a newly developed method was excellent. The creation of wide coaptation zone with complete cleft closure up to chordal attachment might be effective to prevent late LAVVR.

P1225 - RESTRICTIVE ENLARGEMENT OF PULMONARY ANNULUS AT SURGICAL REPAIR OF TETRALOGY OF FALLOT A COMPARATIVE 10 YEAR FOLLOW UP STUDY

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Background: Since 1996 our center follows a uniform strategy of restrictive enlargement of the pulmonary annulus (PV) at surgical repair of tetralogy of Fallot (ToF). A transanular patch (TAP) is only used if the z-score of the PV is < -2. We sought to determine whether this strategy leads to reduction of pulmonary insufficiency (PI) and re-operation rate in the long-term compared to a nationwide contemporary cohort that has not been operated using a uniform strategy.
Methods: 87 ToF patients (repair 1996-2006) were included in the study (Group 1). Echocardiographic, ECG and cardiac MRI data were collected. 172 ToF patients from the Competence Network for Congenital Heart Disease served as the controls (Group 2).
Results: Follow Up time since repair was not different between groups (12.9 (7.7-18.8) vs. 13.1 (5.2-16.9) years, $p = 0.9$) while TAP rate was significantly lower in Group 1 (32.2% vs. 62.8%, $p < 0.001$). In Group 1 more patients were in NYHA class 1 ($p < 0.01$), QRS was narrower (132 ± 22 ms vs. 144 ± 21 ms, $p < 0.001$), moderate or severe PI was less common ($p < 0.001$), CMR derived volumes were smaller (EDVi: 103 ± 21 vs. 120 ± 30 ml/m^2, $p < 0.001$; ESVi: 52 ± 15 vs. 59 ± 19 ml/m^2, $p = 0.03$) and regurgitation fraction was lower (21 ± 14 vs. 29 ± 16%, $p < 0.01$). 10 year freedom from re-operation of the PV was significantly higher in our cohort (92% vs. 87%, $p = 0.015$).
Conclusion: In long-term follow up restrictive enlargement of the PV reduces the rate of pulmonary insufficiency and limits the amount of right heart dilatation. The re-operation rate of the PV is significantly lower compared to a contemporary cohort.

P1231 - CHALLENGE IN MANAGEMENT OF PATIENTS WITH FUNCTIONALLY UNIVENTRICULAR HEART IN INDONESIA

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Management of patients with functionally univentricular heart has evolved during recent years. Usually patient underwent stages operation to achieve "normal" functional heart towards Fontan circulation. In our countries with large population and scattered around many islands, it is still difficult to manage patients with heart diseases. Especially for stages operation, while economy still a big problem for young family. Centre performing complex procedures on a regular basis is very few in Indonesia including Fontan Procedures. Harapan Kita is the biggest center for doing pediatric cardiac center in Indonesia. From 1998 to 2016 there were 661 childrens underwent BCPS operation, meanwhile only 171 (26%) of those patients had Fontan operations. There are a few reasons why only small number of patients had Fontan completion. Previously, government did not give fully support for operation, but for recent years it is covered by government insurance although only for selective population. That is why, the number of Fontan operation is increasing for the last four years. But some patients had long time to wait for Fontan completion, thus it will compromise the condition of the heart to have Fontan operation. Careful selection of patients and optimal timing of intervention remain cornerstones of successful outcomes. But only relatively few patients will fulfill all of Fontan criteria. While, however, the 'limits' of Fontan inclusion criteria continue to be challenged, it remains the case that preoperatively impaired ventricular function and elevated mean pulmonary arterial pressure (greater than 15 to 20 mmHg) still have an adverse impact on surgical outcomes in contemporary series.

P1248 - POLYMERIC BIOABSORBABLE VASCULAR GRAFT IN MODIFIED FONTAN PROCEDURE – TWO YEAR FOLLOW UP

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Background/Hypothesis: The bioabsorbable graft material is designed to allow patient's own cells to infiltrate and replace the material by a process called Endogenous Tissue Restoration (ETR) leading to natural tissue growth. After implantation the graft is gradually replaced with the native tissue, developing into a fully functioning blood vessel. This study evaluated safety and performance of a novel bioabsorbable vascular graft in pediatric patients with univentricular congenital malformation, undergoing hemodynamic correction with an extracardiac cavopulmonary conduit.
Materials and Methods: Five patients with single ventricle congenital malformation have been enrolled in the study from October 2013 to February 2014. In all patients the bi-directional Glenn anastomosis have been previously performed. The patients' age at the time of implantation were ranged 4–12 years while 60% ($n = 3$) were males. Total follow-up was 26–31 months with scheduled visits at 1, 3, 6, 9 and 12 months after surgery for the first year and yearly thereafter. The device performance evaluation has been performed by transthoracic ultrasound, and optional CT-scan and MRI were performed at several timepoints in all patients.
Results: All 5 patients have been successfully recovered from the procedure and completed 24 months follow-up. No device related adverse events were reported to date. Ultrasound and MRI studies have demonstrated anatomical (conduit dimensions) and functional (blood flow velocity and pattern, absence of thrombosis) stability of the grafts in all patients at up to 31 months.
Conclusions: The study device demonstrated an adequate hemodynamic performance at up to 31 months follow-up. The clinical study outcomes suggest that this bioabsorbable polymer technology has the potential to improve cardiac and vascular surgical procedures by reducing implant-related complications. Longer follow-up, however is needed to fully assess effectiveness of bioabsorbable vascular grafts. This represents the first step towards development of more complex bioabsorbable devices such as heart valves.

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Table. Postoperative conduit diameter (A, mm) and maximal blood flow velocity (B, cm/sec) assessed at the mid-level of the graft at 12 and 24 months follow-up.

Patient	01-001		01-002		01-003		01-004		01-005	
F/u, mo	12	24	12	24	12	24	12	24	12	24
A	20	19.7	16	18	18	19.5	18.2	19.7	n/a	18
B	25	38	32	75	13.5	25	16	25	n/a	35

P1249 - HYBRID PULMONARY VALVE IMPLANTATION IN TETRALOGY OF FALLOT

María-Teresa González-lópez[1], Ramón Pérez-Caballero-Martínez[1], Fernando Ballesteros-Tejerizo[2], Alejandro Rodríguez-Ogando[2], José-luis Zunzunegui-Martínez[2], Ana-María Pita-Fernández[1], Juan-Miguel Gil-Jaurena[1]
Gregorio Marañón Hospital, Pediatric Cardiac Surgery, Madrid-Spain[1]; Gregorio Marañón Hospital, Pediatric Cardiology, Madrid-Spain[2]

Background: Transannular patch repair of tetralogy of Fallot (TOF) often results in significant right ventricular outflow tract (RVOT) dilation and distortion, along with free pulmonary regurgitation (PR). Pulmonary valve replacement (PVR) is indicated in this group of patients at any point of the follow-up. The surgical and trans-catheter options of PVR continue to expand, but hybrid approaches have not been described in this scenario. We present two cases of a novel approach of hybrid PVR and early outcomes.
Patients/Methods: CASE 1. Male, 2.5 year-old. He was referred to our center following complete TOF transannular repair (6 months of age). A residual ventricular septal defect (VSD), moderate pulmonary stenosis and nodal rhythm were found. He underwent VSD closure, left pulmonary artery (PA) patching, implantation of Melody valve (expanded to 16 mm; four interrupted sutures) and definitive epicardial leads. Cardiopulmonary bypass time 155 min; aortic cross clamping time 58 min. CASE 2. Female, 2 year-old. She underwent palliation procedure (5 months of age): transannular patch and right ventricular (RV) hypertrophy resection. The VSD was left open due to severe hypoplasia of PA branches). Two months later, left PA stent was implanted. One year later, normal-sized PA branches were noted. She underwent VSD closure (pericardial patch) and implantation of Melody valve (expanded to 14 mm). Cardiopulmonary bypass time 154 min; aortic cross clamping time 56 min.
Results: Both patients had an uneventful postoperative course. At follow-up (12 + /-2 months), they remain asymptomatic with optimal functional class. Last echocardiography has showed good RV function, normal PV function and good-sized PA branches.
Conclusions: This novel approach may offer potential benefit in treating complex TOF patients. It provides a reasonable alternative to surgical PVR when any additional surgical procedure is needed at an early age, allowing a more adequate matching to the RVOT size as well as subsequent balloon expansions.

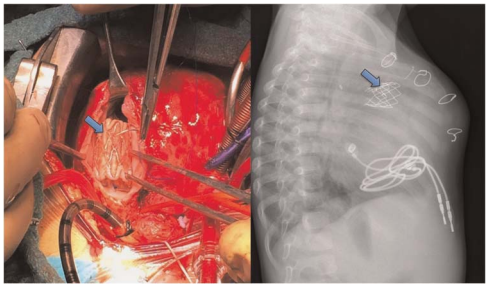

Figure.

P1258 - EUGENY MESHALKIN (1916–1997) THE PIONEER OF THE WORLD HEART SURGERY (THE FIRST IN THE WORLD CLINICAL SUCCESSFUL CAVAPULMONARY CONNECTION) – IN MEMORIAM

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Gruppo San Donato, Cardiac Surgery Department, Milan-Italy[1]; Siberian Federal Biomedical Research Center, Congenital Heart Diseases Department, Novosibirsk-Russia[2]

Last year it was 100th Anniversary of Eugeny Meshalkin, the pioneer of the world heart surgery. He graduated from high medical school in Moscow just before the Second World War. He was drafted into the army as a military front surgeon for 5 years. In 1946 Meshalkin entered to a clinic in Moscow headed by Aleksandr Bakulev. This clinic became the first center of cardio-vascular surgery in Russia. In 1952 Meshalkin became a chief of the division of cardio-vascular surgery. He performed more than 200 cardio-vascular operations a year. On 3 April 1956 Meshalkin did the first successful operation of cavopulmonary anastomosis for a 15 years old patient with tetralogy of Fallot. He published a paper in the Russian journal "Experimental Surgery" (N 6, 1956) which summarized the experience of 24 operations of cava - pulmonary anastomosis performed from April to October 1956. Three patients died. Meshalkin described the technique of the operation in detail. Twenty three anastomosis were "end to end" and one was end of pulmonary artery to side of vena cava. Meshalkin was the first in the world who used mechanical suture for dividing vena cava. He performed postoperative angiography of the shunt. Only two years later on February 25, 1958 William Glenn was the first in North America to perform a successful cavopulmonary shunt, and it became known by his name. Francis Robicsek wrote in his paper "An epitaph for cavopulmonary anastomosis" (the Annals of Thoracic Surgery vol 34, N2, 1982) that the patient Meshalkin operated on April 3, 1956 undoubtedly represents the first successful clinical case of cavopulmonary anastomosis reported in surgical literature. Harris Shumacker, who dedicated a lot of works to developing this operation, confirmed the priority of Meshalkin in his book "Evolution of Cardiac surgery" in 1992.

P1259 - WHEN THE CHILD BECOMES A YOUNG ADULT UNCOMMON SCENARIOS FOR AORTIC VALVE REPAIR

María-Teresa González-López[1], Ana-María Pita-Fernández[1], Ramón Pérez-Caballero-Martínez[1], Alejandro Rodríguez-Ogando[2], Constancio Medrano-López[2], Juan-Miguel Gil-Jaurena[1]
Gregorio Marañón Hospital, Pediatric Cardiac Surgery, Madrid-Spain[1]; Gregorio Marañón Hospital, Pediatric Cardiology, Madrid-Spain[2]

Background: Aortic regurgitation (AR) can be developed over the time following surgical repair of several congenital heart defects. Aortic valve repair needs to be highly considered in this group of young patients. We present 2 uncommon scenarios of aortic valve repair and short-term results.
Patients and Methods: CASE #1. Female, 17 year-old. She underwent surgical closure of subarterial ventricular septal defect (VSD) (Laubry-Pezzi syndrome) at 4 years of age. At long-term follow-up, significative intolerance to exercise was detected. Moderate-to-severe AR and small residual VSD was noted. Left ventricular ejection fraction (LVEF) was 60%. On the operative field, severe fusion of the right and left coronary leaflets with severe retraction of right coronary leaflet were noted. Commissurotomy and shaving of the left coronary leaflet were performed followed by a complete excision of the right coronary leaflet. A right neo-leaflet

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(equine pericardial patch, glutaraldehyde-free, continuous suture) was created and the VSD was closed. Cardiopulmonary bypass time 104 min; aortic cross clamping time 71 min. CASE #2. Male, 17 year-old. He underwent arterial switch operation (Lecompte maneuver) in the neonatal period due to transposition of the great arteries (TGA). At long-term follow-up, severe AR was detected with preserved LVEF. A valve-sparing procedure (David operation) was performed, following the transection of right pulmonary artery for an optimal exposure of the aortic root. Cardiopulmonary bypass time 187 min; aortic cross clamping time 95 min.
Results: Both patients had an uneventful postoperative course. At follow-up (14 + /-2 months), they remain asymptomatic with optimal functional class. Last echocardiography has showed good LV function with no AR.
Conclusions: A repair-oriented strategy for the aortic valve has excellent short-term results in complex congenital patients. Although these procedures are technically demanding, they remain a promising management option in these uncommon scenarios.

P1268 - BEATING HEART AORTIC ARCH REPAIR

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Introduction: The strategies for aortic arch repair (AAR) in neonates/infants have evolved notoriously. Simultaneous cerebro-myocardial perfusion (beating-heart technique) is the most recent modification, although not widely performed. We present our preliminary results by using this strategy.
Methods: Since 2013 we included this technique as the standard approach, gathering 44 patients in four years. Median age 50 + /-27 days (range 1-270); median weight 3.7 + -1.4 kg (range 2.1-7). Hypoplastic aortic arch (HAA) was associated with cor triatriatum (3), ventricular septal defect (VSD,10); interrupted aortic arch (3), transposition of great arteries (5), atrial septal defect (ASD, 3) partial atrioventricular septal defect (AVSD, 1), hypoplastic left heart syndrom (HLHS, 7), double-outlet right ventricle (2), severe aortic stenosis (2) and as isolated defect (8). Coronary perfusion (25 °C) was maintained through a cardioplegia delivery system Y-connected to the aortic cannula. Hence, heart-beating AAR was performed. When intracardiac repair was required, antegrade cardioplegia was delivered via the same catheter used for myocardial perfusión, simply switching the three-way stopcock.
Results: 32 patients had intracardiac lesions: left atrium membrane resection (3), VSD closure (9), arterial switch operation (5), ASD closure (3), partial AVSD repair (1), Norwood-Sano procedure (7), comprehensive-procedure following hybrid-approach (2) and aortic commisurotomy (2). Average cardiopulmonary bypass time 168 + /-73 minutes (range 93-312). Myocardial ischemia time median 30 + /-11, (range 0-160). The heart was not arrested in12 patients: 8 isolated hypoplastic arch, 2 Glenn, 1 Banding, 1 perventricular VSD closure. Selective cerebral perfusion median 35 + /-9 minutes (range 18-53). No cases of operative mortality were detected. No neurologic adverse events occurred. 30-day mortality was 4.5% (n = 2, following extra-corporeal-membrane-oxygenation).
Conclusions: Selective/independent cerebro-myocardial perfusion in aortic arch pathology is a safe/feasible technique, with low rates of adverse events. Heart-beating AAR should be recommended due to the reduction of the myocardial ischemia time, although comparative results with classical techniques need to be addressed.

P1269 - SURGERY OVER STENTS. NEW CHALLENGES TO FACE

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Objectives: Complex cases undergo step surgical and percutaneous procedures, including stent deployment. Concerns arise on stent removal at latest surgery. We describe our experience in this issue.
Methods: 83 stents in 68 patients were partial or totally removed at surgery. Univentricular heart was diagnosed in 30 patients. Stents were previously deployed in: ductus (14), right ventricle outflow tract (RVOT, 20), atrial septal defect (ASD, 6), right pulmonary artery (RPA, 9), left pulmonary artery (LPA, 23), inferior vena cava (IVC) 5, superior vena cava (SVC, 3) ascending aorta (AAo, 2) and pulmonary veins (1). Surgical procedures performed: 14 transplants, 10 Fontan, 4 Glenn, 2 comprehensive repair (Norwood + Glenn), 1 Glenn take-down, 13 conduit replacement, 8 Fallot, 6 Rastelli, 1 ventricular septal defect closure (VSD), 1 iatrogenic aorto-pulmonary window, 1 Ross-Konno, 1 double switch, 1 hypoplastic aortic arch and 1 unifocalization.
Results: Ten ductal stents were clipped. Nineteen stents in RVOT, six ones in ASD, five in RPA, thirteen in the LPA, three in IVC and one in the ascending aorta were completely removed. Five stents in RPA, three in SVC, one in AAo and thirteen in the LPA were split and partially retrieved. Handling the stents in ductus, RVOT and ASD was fairly seamless. On the contrary, stent removal in the ductus (for the two comprehensive cases), RPA, LPA, SVC, IVC, aorta or pulmonary veins required short periods of deep hypothermia with circulatory arrest, adding length and morbidity to the procedure.
Conclusions: surgery over stents is increasing in complex, step procedures. Univentricular hearts are most prevalent. Congenital Transplant surgery faces new challenges. Stent removal at the time of surgery may require deep hypothermic circulatory arrest.

P1274 - A RARE CASE OF SITUS INVERSUS TOTALIS VENTRICULAR SEPTAL DEFECT WITH INFUNDIBULAR PULMONARY STENOSIS

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Introduction: 4 years old male child (12 kg) was admitted with complaints of Breathlessness on exertion for the past 6 months. On Investigation, he was found to have Congenital Heart Disease, Situs Inversus Totalis, Perimembranous VSD with Severe Infundibular PS.
Surgical Strategy: We took certain precautions for ease of doing surgery in Situs Inversus Totalis like - Positioning the patient in the centre of the operating table. - Taking the Arterial and Venous lines little high up for the ease of the movement of the surgeon toward the left side of the patient. - Canulation from right side of the patient. - Main procedure after arresting the heart from left side of the patient. - Decannulation and Closure from the right side of the patient.
Procedure Performed: Trans atrial- Trans Pulmonary Infundibular muscle bundle resection and closure of VSD with 0.6mm PTFE patch.
Conclusion: Cardiac surgery in Situs Inversus Totalis is no different from others, provided we take certain precautions and follow few simple surgical strategies..

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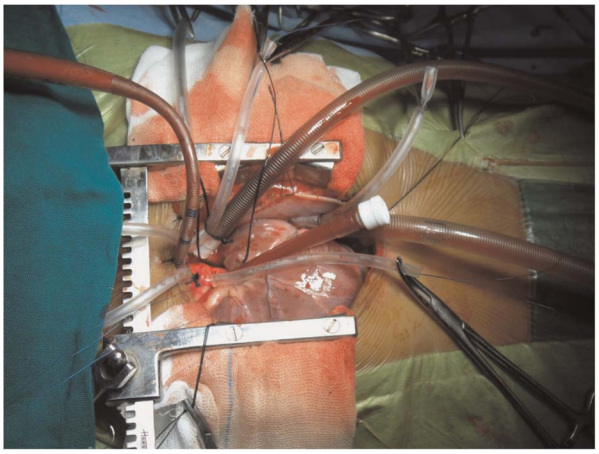

Figure 1.

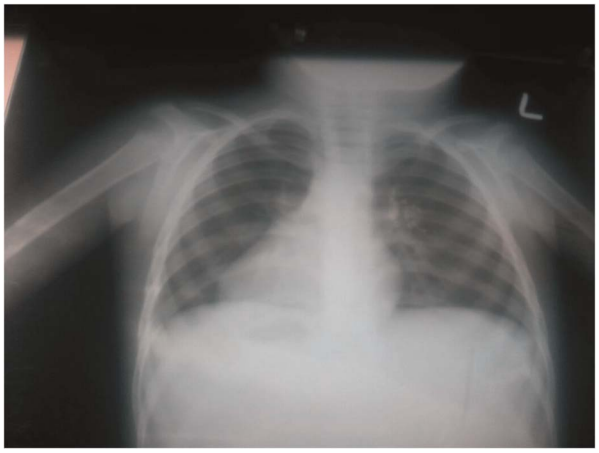

Figure 2.

P1280 - OPTIMAL TIMING FOR SUREGERY ON AORTIC VALVE INSUFFICIENCY IN PEDIATRIC PATIENTS INSIGHT ABOUT PREOPERATIVE INDICES TO PREDICT POSTOPERATIVE RECOVERY OF LEFT VENTRICULAR GEOMETRY

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Background: Though indexed end-systolic LV dimension (ESDI) < 25 mm/m2 is reported to be an optimal timing to maintain LV function after surgery for chronic aortic valve insufficiency (AI) in adults, we have paucity of data describing the optimal timing for aortic valve repair (AOVR) on pediatric AI.
Objectives: The aim of this study is to establish the cut-off values of ESDI and indexed end-diastolic LV dimension (EDDI) to determine the optimal timing for AOVR on pediatric AI.
Methods: 36 consecutive pediatric AI (moderate 8, severe 28) who had biventricular physiology and underwent AOVR between January 2001 and December 2015 were enrolled. 24 were male. Age at AOVR was 10.4 + /- 4.7 (mean + /- S.D.). All patients didn't receive any intervention within 1.5 year before AOVR. Aortic valve replacements were done on 16, Ross procedure on 11, and aortic valve plasty on 9. When ESDI became < 25 and EDDI <40 after AOVR, they are classified as normalized group (N). The others are considered as dysfunctional group (D). Preoperative ESDI and EDDI were compared between two groups.
Results: Preoperative ESDI is significantly smaller in N than D (28.1 + /- 6.5 vs 41.9 + /- 15.4, p = 0.002), and the cut-off value differentiating two groups is 39.9 (area under curve [AUC]: 0.811). Preoperative EDDI is also significantly smaller in N than D (46.1 + /- 9.6 vs 66.1 + /- 18.3, p = 0.006) and cut-off is 51.6 (AUC: 0.869). Preoperative ejection fraction was not different between the groups. By multiple logistic regression analysis, preoperative ESDI <40 predicts normalization of LV geometry after surgery (N = 88% [22/ 25], D = 27% [3/ 11], odds ratio = 1.42 [95% confidential interval; 1.07-2.13], p = 0.03).
Conclusion: Preoperative ESDI <40 would predicts improvement of LV geometry after surgery for pediatric AI.

P1281 - STAGED REPAIR OF PULMONARY ATRESIA VENTRICAL SEPTAL DEFECT AND MAJOR AORTO PULMONARY COLLATERAL ARTERIES WITH SEVERE HYPOPLASIA OR ABSENCE OF PULMONARY ARTERIES

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Background: Surgical management of pulmonary atresia, ventrical septal defect and major aorto-pulmonary collateral arteries with severe hypoplastic or absent pulmonary arteries is still a challenge. We evaluated the results of staged surgical treatment in such challenging patients.
Methods: From 2000 to 2015, 32 consecutive patients with pulmonary atresia, ventrical septal defect and major aorto-pulmonary collateral arteries were surgically treated. Primary complete repair was done only in 4 patients, another underwent staged procedures. Treatment of 28 staged patients began with systemic-to-pulmonary shunt only, unifocalization and systemic-to-pulmonary shunt or palliative right ventricular outflow tract reconstruction and depending on the distribution of pulmonary blood flow. Among all staged patients, 22 had severe hypoplastic pulmonary arteries (median Nakata index 46,7 mm/m2, range 13,1-135,4 mm/m2), and 6 had inconfluence or absence of pulmonary arteries.
Results: 10 staged patients (36%) subsequently underwent complete repair with ventricular septal defect closure (median 2,5 surgical procedures, range 2-4). The median of the pulmonary artery pressure to the systemic blood pressure ratio at the final stage of repair was 0.66 (range 0.44-1.00). Duration of the full surgical course ranged from 6,6 to 69,2 months (median 15,7 months). Overall hospital mortality in staged group was 14,3% (3 patients died after staged procedures and 1 – after complete repair). Subsequently, 3 patients underwent interventional catheterizations (dilation or/and stenting of pulmonary arteries), and right ventricle – pulmonary artery conduit was replaced in 1 case.
Conclusion: Staged surgical approach can be used in pulmonary atresia, ventricular septal defect with severe hypoplastic or absent pulmonary artery and major aortopulmonary collateral arteries with acceptable results.

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P1282 - OUTCOMES OF AORTIC VALVE REPAIR IN NEONATES AND INFANTS

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Background: To investigate the outcomes in neonates and infants who underwent aortic valve repair.
Materials and Methods: Five patients, aged <1 year, who underwent aortic valve repair between 2012 and 2016, were evaluated. The primary diseases were critical aortic stenosis in 2 cases, aortic stenosis + coarctation of the aorta in 1, left isomerism + aortic atresia in 1, and truncus arteriosus in 1. The median age and body weight at the time of surgery were 40 days (range: 21–202 days) and 3.3 kg (range: 2.6–5.2 kg), respectively. In all cases, aortic valve repair was conducted after bilateral pulmonary artery banding as the initial surgery; in 3, transcatheter aortic valvotomy was performed before aortic valve repair. The surgical procedures for aortic valve repair were leaflet extension + commissure plication in 2 cases, closure of the aortic valve leaflet hiatus + commissure plication in 1, commissurotomy in 1, and truncal valve remodeling in 1.
Results: Follow-up evaluations were performed between 1 and 23 months post-surgery (median: 19 months). One patient died 1 month after surgery from an unrelated condition; the remaining 4 survived. The mean (± SD) flow velocity in the aortic valve was reduced from 3.4 ± 1.1 m/s to 2.8 ± 0.7 m/s after surgery. Two patients required further aortic valve repairs. One patient required suture of the commissure aortic valve 2 months after the initial repair. In another case, the transcatheter aortic valvotomy was performed twice, and Ross-Konno procedure was performed at 8 months after the initial repair. At the latest examination, the left ventricular function was maintained, and B-type natriuretic peptide decreased (pre-surgery: 965 ± 704 pg/ml, post-surgery: 87 ± 103 pg/ml).
Conclusions: The outcomes of the aortic valve repairs were fairly favorable. The results suggest that additional transcatheter aortic valvotomy and/or valve repair after the initial repair was useful to increase the options for future procedures, including Ross-Konno procedure and aortic valve replacement.

P1291 - MULTI DISCIPLINARY FOUR PRONGED TEAM COMMUNICATION THROUGHOUT THE OPERATIVE DAY FOSTERS BETTER UNDERSTANDING FOR COMPLEX PATIENTS AND IMPROVES OUTCOMES

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Background: A team that communicates openly, defines patient goals, and utilizes a consistent management approach; can reduce care variability, errors and improve outcomes. Well-executed transitions of care improve patient safety and outcomes. Using several tools can help drive understanding. The four important processes we use are: (1) Pre-brief – outlines the conduct of the surgical procedure and expectations, (2) Pre-Op Huddle – Pre-defined worksheet with goals of the Surgeon, Anesthesia, Perfusion, and ICU, performed the morning of surgery, (3) Post-Op-Debrief (POD) – Review the Huddle sheet in the operating room, and define any changes or discrepancies, and (4) Team handoff performed in the ICU.
Method: In 5/1/15, we enhanced our process to include intensive care team in the Huddle and post-huddle process. The Huddle sheet was changed and the huddle was moved to the PICU conference room. The ICU team ran the post-huddle starting in 5/1/15. We compared the entire process (1) Pre-brief, (2) Huddle, (3) Post-Huddle and (4) handoff; pre and post changes. Real-time audit of all forms, with observations of huddles by independent observers and using event monitoring to determine any clinically significant events.
Results: Two surgical groups November 2014 through April 2015 (n = 19) and May 2015 through April 2016 (n = 26). In all patients there was complete documentation of allergies, patient weight, anesthesia plan, and perfusion plan. Surgical plan (100% vs. 96%) and nursing plan (95% vs. 92%) was almost always complete.
Conclusions: A team approach to care had a tremendous effect on patient outcomes, by improving communication of the entire team. We found there was a significant reduction of overall issues immediately in the ICU. By having the ICU team more involved (moving the Huddle to the ICU and having the ICU team run the post-huddle) there was a greater acceptance.

Table 1.

	Group 1	Group 2	p-value
Dosing Weight Documented	0%	50%	<0.001
Post-Op Debrief Completed	5%	92%	<0.001
ECMO Plan Determined in post-Op Debrief	5%	88%	<0.001
ICU Plan Defined in the post-Op Debrief	11%	77%	<0.001
Significant Clinical Disturbances	2.3 ± 1.7	0.7 ± 1.6	0.003

P1295 - EVALUATION OF PERICARDIAL MITRAL VALVE RECONSTRUCTION FOR PEDIATRIC PATIENTS

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Objective: Mitral valve or left sided atrioventricular valve is often involved in structural congenital heart diseases in pediatric patients. The size of commercially available prosthetic valves limits the usage of valve replacement in this group of patients. We hypothesize that pericardial reconstruction of mitral valve with newly developed mold by our group is a feasible option.
Methods: The finite element computer models of a reconstructed mitral valve were created. The leaflet coaptation area and stress distribution on the leaflets were analyzed. Bovine pericardium was prepared based on the computer model. It was hand-sewn to the pigs' hearts as well as the silicon test-rig. Valve motions were observed and recorded with the video camera.
Results: Von Mises stress on the leaflets ranged from 0.47 to 0.85 MPa. The highest stress distribution was observed in the commissure at the chordae. From the video recording on the pigs'

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hearts, the mitral valve closed rapidly with native-like coaptation and revealed small amount of leakage. The silicon test-rig confirmed similar findings.
Conclusions: The computer model of reconstructed mitral valve showed acceptable performance. Pericardial reconstruction of mitral valve showed similar appearance and performance to the native valves. Although further evaluation is necessary, this has a potential to be a feasible option for pediatric patients.

P1301 - SURGICAL REPAIR OF SUPRAVALVULAR AORTIC STENOSIS SURGICAL RESULTS AND 44 YEARS FOLLOW UP

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Objective: Supravalvular aortic stenosis is defined as left ventricular outflow tract obstruction distal to the aortic valve. Hospital results and long-term follow up of the surgical repair were analyzed and different technical options were compared.
Methods: Between 1972 and 2016, 34 patients younger than 18 years were operated in our department; 70% of whom were male, ranging in age from 0.5 to 15 years (average age, 7 ± 4 years). 23 patients (67.6%) had Williams-Beuren syndrome. Preoperatively, 79.4% patients were asymptomatic. Among the 34 patients, 30 (88.2%) had localized type and 4 diffuse type obstruction. Preoperative peak gradient was 88 ± 33 mmHg and mean gradient was 49 ± 23 mmHg. Left ventricular ejection fraction (LVEF) was conserved in all of them.
Results: Single-sinus repair was performed in 13 patients, two-sinus in 16, and three-sinus in 1. In the remaining four patients, an aortoplasty of a diffuse form was carried out. There was one hospital death. Within the follow-up period ranging from 0.5 to 44 years (medium 21 ± 12 years) one late death happened in the 42nd year and 9 patients had one additional operation (27.3%). Currently the peak gradient is 20 ± 12 mmHg and mean gradient is 13 ± 16 mmHg. LVEF was preserved in 94% (32). There was no statistical significance ($p = 0.17$) in additional operation between the single-sinus technique and the others.
Conclusion: Hospital death and late survival were excellent, with a small numbers of additional operations, no association was found depending on the surgical technique used. Surgical indication should be considered in asymptomatic patients with peak gradients > 50 mm Hg or average gradients > 30 mm Hg with ventricular hypertrophy.

P1303 - EARLY OUTCOMES OF THE ARTERIAL SWITCH OPERATION IN CHILDREN OF MORE THAN 1MONTH OF AGE

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Objectives: Though results of arterial switch operation (ASO) in children presenting early have been encouraging, late presenters continue to pose a challenge. This study aims therefore aims to evaluate out the early results ASO in such children more than 1month of age.
Methods: Records of 488 patients undergoing ASO from January 01, 2005 till April 30, 2016 at All India Institute Of Medical Sciences (AIIMS), New Delhi, India, were analysed. They were divided in 3 groups, Group A (TGA with intact ventricular septum (IVS)), Group B (TGA with VSD) and Group C (Taussig- Bing Anomaly).
Results: Of the 488 children reviewed, records of 470 were available. A total of 268 (57%) had associated VSD, 183 (37.9%) were had intact IVS and 24 (5.1%) presented with DORV. Other associated conditions found were left ventricular outflow obstruction (10/470, 2.1%), coactation of aorta (3/470, 0.6%), right ventricular outflow obstruction (3/470, 0.6%), pulmonary stenosos (2/470, 0.4%), aortopulmonary window (1/470, 0.2%), total anomalous pulmonary venous drainage (1/470, 0.2%) and hypoplastic aortic arch (1/470, 0.2%). Overall early hospital was 11.7% (55/470). In Group A, B & C early hospital mortality was 10.7% (19/178), 13.1% (35/268) and 4.2% (1/24) respectively. ECMO was instituted in 8.9% (42/470) patients. Requirement of ECMO was significantly higher ($p < 0.005$) for children having IVS (38/42 i.e, 90.5%) as against children having VSD (4/42 i.e, 9.5%).. On detailed evaluation of the possible incriminating risk factor for deaths, in such children, presence of associated conditions such as arch abnormalities, presence of CoA, pre-operative infections were found.
Conclusion: In a developing countries, late presentation of TGA is not uncommon. However acceptable results can be obtained in these patients.

P1305 - A RANDOMISED CONTROLLED TRIAL COMPARING RIGHT VENTRICULAR FUNCTION BETWEEN TRANS RIGHT ATRIAL VERSUS TRANS RIGHT ATRIAL RIGHT VENTRICULAR APPROACH FOR TETRALOGY OF FALLOT REPAIR

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Background: We compared the immediate pre and postoperative right ventricular functional status byTrans Annular Plane Systolic Exertion (TAPSE) between trans-right atrial (RA) versus trans-right atrial/right ventricular (RA/RV) approach for TOF repair.
Methods: 50 consecutive paediatric patients between ages of 01 year to 15 years undergoing intracardiac repair of TOF between July 2015 and June 2016 were randomized in two groups based on the approach for repair, namely; trans- RA and trans-RA/RV.
Results: Both groups were well matched in terms of age, body surface area and preoperative saturation, bypass time and aortic cross clamp times, ionotropic score, post-operative ICU stay and hospital stay. However Trans RA/RV group had significant mediastinal drain (169 ± 163 ml vs 90.6 ± 58.7 ml, $p < 0.05$), pleural effusions (8 vs 2 patients, $p < 0.05$) but had better relief or RVOT gradient. There was no difference in incidence of post-operative arrhythmias in either group till the first month of follow-up. Preoperative TAPSE for trans RA and trans RA/RV were similar (1.49 ± 0.29 vs 1.66 ± 0.34, $p > 0.05$) and so was the post-operative TAPSE at discharge (1.52 ± 0.30 vs 1.43 ± 0.32, $p > 0.05$) and at one month post operatively (1.6 ± 0.27 vs 1.43 ± 0.032, $p > 0.05$).
Conclusions: Both trans-RA and trans RA/RV approaches provide a safe palliation for patients with TOF. The right ventriculotomy should not be avoided if required for adequate infundibular resection and can be safely done without fear of producing excessive arrhythmias and early right ventricular dysfunction.

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P1310 - CONGENITALLY CORRECTED TRANSPOSITION INTERMEDIATE AND LONG TERM OUTCOME OF DIFFERENT SURGICAL TECHNIQUES

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Objectives: To assess the outcome of different surgical techniques in patients with congenitally corrected transposition of the great arteries (CCTGA).
Material and Methods: 66 out of 133 patients diagnosed with CCTGA underwent surgery between January 1998 and February 2016. Group 1 (Anatomic repair): 20 patients (25.3%); mean age: 44.6 months. Surgical techniques: Double switch in 10 patients; Senning /Mustard + Rastelli in 8 patients; Hemimustard + Switch, 1 patient; Hemimustard + Nikaidoh, 1 patient. Mean follow-up time: 56 ± 42 months. Group 2 (Traditional surgery): 46 patients (58.2%); mean age: 58.17 months. Surgical techniques: ventricular septum defect (VSD) closure + Left ventricle conduit-pulmonary artery (LV-PA), 33 patients. VSD closure, 10 patients. Tricuspid valve replacement, 3 patients. Mean follow-up time: 104 ± 95 months. The variables evaluated were: tricuspid insufficiency, ventricular dysfunction, complete AV block, aortic insufficiency and reoperations.
Results: Group 1: Improvement in tricuspid regurgitation: 42.8% of cases during follow-up. Ventricular dysfunction: 47% at immediate post-operative time; 50% of them recovered at late follow-up. Two patients developed AV block (with permanent pacemaker). Mild aortic insufficiency: 60%; reoperations, 29.4%. Early mortality rate: 15%; 120-month survival rate: 84%. Group 2: Tricuspid regurgitation: 41%; ventricular dysfunction:28.26%. AV block: 26%: 5 at immediate post-operative time and 9 at late follow-up. Aortic insufficiency: 19.5%. Reoperations: 31.4%. Early mortality rate: 19.5%; 120-month survival rate: 79%.
Conclusions: Outcome of traditional surgery is satisfactory, although there is a tendency for progression towards tricuspid insufficiency and AV block with requirement of permanent pacemaker. Anatomic repair shows improvement of tricuspid regurgitation during follow-up, with low rate of complete AV block, but a greater risk of ventricular dysfunction at immediate post-operative time, and intermediate and long-term mild aortic insufficiency. Reoperation rate was similar in both groups.

P1314 - EARLY RESULTS OF MINITHORACOTOMY USE FOR DUCTUS ARTERIOSUS CLOSURE IN VERY LOW BIRTH WEIGHT PRETERM INFANTS

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Background: Ductus arteriosus' (DA) functional closure occurs 12-15 hours after birth, and permanent closure 5-7 days later. Studies show that 58,8% of very low birth weight neonates have patent ductus arteriosus (PDA), and if left open longer than usual it can cause severe complications such as pulmonary edema, necrotizing enterocolitis, heart and renal failure, systemic hypotension, and cerebral blood flow alterations. Recent studies suggest initial pharmacological treatment, and if unresolved, proceed to surgical intervention, but other findings point out lower immediate mortality of patients treated surgically right after PDA diagnosis. Our objective is to analyze the immediate mortality of infants submitted to surgical closure of the DA.
Material and Methods: From March 2001 to December 2015 the Esperança Hospital of Recife's medical records of 32 very low birth weight preterm neonates with PDA were analyzed. Corrected gestational age varied from 24 to 36 weeks (m = 29 weeks) and weight ranged from 400 to 1,000 g (m = 743,1 g). Surgery performed under general anesthesia, hemodynamic monitorization with noninvasive arterial pressure, pulse oximetry and continuous EKG. The DAs were resected through 2 cm minithoracotomies on the 4th left posterior intercostal space. Metallic clips used according to the DA's size (LT300 or LT400 Ethicon®). After clipping, there was an improvement in the systemic arterial pressure and disappearance of the fremitus.
Results: After the surgical procedure, only one patient (3,1%) needed pulmonary drainage due to a pneumothorax. None of the patients presented surgical site infection. Immediate surgical mortality was of one patient (3,1%).
Conclusion: Although surgical procedures may be high risk for very low birth weight premature infants, DA closure through minithoracotomy had minor morbidity and mortality. Future studies are needed to analyze late results.

P1316 - FONTAN OPERATION FOR 150 CONSECUTIVE CHILDREN WITH HYPOPLASTIC LEFT HEART SYNDROME WITHOUT EARLY MORTALITY

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Background: The operative results of the multistage reconstructive surgery for children with hypoplastic left heart syndrome (HLHS) are improving in the recent years and more patients reach the last stage surgery. The goal of the study was to analyze the hospital results of the Fontan operation for children with HLHS.
Material and Methods: Between September 2007 and August 2016, 150 children in the mean age of 37.6 ± 15.6 months and weight of 12.4 ± 1.9 kg with HLHS underwent Fontan operation, and were operated on by the same surgical team in two institutions. 112 patients after bidirectional Glenn anastomosis underwent extracardiac conduit (EC) on beating heart (only 8(7.1%) children required aortic cross-clamp for intracardiac procedures), and 38 children after hemi-Fontan operation underwent lateral atrial tunnel (LT) Fontan procedure. The medical records were retrospectively analyzed.
Results: The hospital survival was 100%. The mean intubation time was 7.2 ± 6.2 hours, mean stay in the intensive care unit was 3.9 ± 9.2 days and the mean hospital stay was 17.7 ± 13.2 days. The mean cardio-pulmonary bypass time (EC: 45.1 ± 14.0 vs. LT: 62.1 ± 17.1 min; $p < 0.001$), mean aortic cross-clamping time if needed (EC: 24.4 ± 6.7 vs. LT: 32.2 ± 7.6 min; $p = 0.016$) and mean intubation time (EC: 5.1 ± 7.4 vs. LT: 9.3 ± 5.9 h; $p = 0.002$) were shorter in the EC group. Children after hemi-Fontan procedure required during Fontan operation significantly

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more often tricuspid valve repair or replacement in comparison with children after Glenn anastomosis (LT: 9(23.7%) vs. EC: 8 (7.1%); p = 0.013).
Conclusions: Fontan operation in children with HLHS can be performed nowadays without hospital mortality and with low morbidity rate, regardless of the operative technique. The extracardiac conduit Fontan operation can be performed with shorter cardio-pulmonary bypass time and without cardioplegic cardiac arrest on beating heart, which can have impact on the myocardial performance in the late follow up.

P1318 - EXTUBATION IN THE OPERATING ROOM FOR CHILDREN AFTER FONTAN OPERATION

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Background: Early extubation is beneficial for the Fontan circulation. The goal of the study was to assess the impact of the extubation on the operating table in comparison with the early extubation in the first hours after Fontan operation (FO) on the early postoperative course.
Materials and Methods: Between 2013 and 2016, 114 children (mean age, 3.8 ± 2.3) with univentricular heart underwent FO in our institution. 60 patients were extubated in the operating room (ORE) and 54 in the intensive care unit (ICUE) in a median time of 195 min (range, 30-515 min) after FO. The medical records (191 parameters) were retrospectively analyzed.
Results: One patient needed a reintubation because of respiratory insufficiency (laryngospasm) (ORE: 1/60 vs. ICUE: 0/54; p = 0.958). The hospital survival was 100% and the mean hospital stay was 17.5 ± 6.8 days, without differences between the groups. The results are presented in the table.
Conclusion: Extubation in the operating room after Fontan operation is feasible, safe and can be routinely performed. The adaptation for the Fontan circulation is faster and needs less therapeutic interventions in children with immediate extubation after surgery.

Table.

	ICUE (N = 54)	ORE (N = 60)	p-value
Heart rate [bpm]*	120.3 ± 5.96	106.5 ± 3.99	<0.001
Blood pressure [mmHg]*			
arterial systolic	93.5 ± 6.43	90.6 ± 2.09	0.024
arterial diastolic	53.6 ± 4.33	49.6 ± 1.38	<0.001
central venous	11.4 ± 1.42	10.4 ± 0.66	0.001
Urine output [ml/kg/h]*	4.8 ± 0.96	3.7 ± 0.78	<0.001
Blood gas analysis 2-3 h after FO			
pH	7.300 ± 0.47	7.364 ± 0.04	<0.001
pCO_2	42.67 ± 7.35	38.25 ± 4.73	0.005
pO_2	86.28 ± 34.20	100.22 ± 40.30	0.048
$SatO_2$	91.58 ± 10.57	93.50 ± 10.46	0.041
Cumulative dopamine support [h]	12.77 ± 11.68	9.75 ± 11.28	0.033
Crystalloid fluid therapy [ml/kg/24 h]	73.76 ± 46.09	54.11 ± 31.19	0.019
Total amount of effusions [ml/kg]			
6 h after FO	11.1 ± 6.72	6.5 ± 3.68	0.024
24 h after FO	31.5 ± 18.13	19.1 ± 9.12	0.001
48 h after FO	49.5 ± 26.10	37.9 ± 17.48	<0.001
Antibiotics therapy [days]	5.77 ± 4.63	4.74 ± 3.36	0.037

*bpm - beats per minute; *mean value of every hour recorded parameters during the first 24 h after FO.

P1321 - BETTER IMMEDIATE RESULTS OF JATENE PROCEDURE BEFORE 15 DAYS OF LIFE

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Background: Transposition of the great vessels (TGV) is the most frequent congenital cyanotic cardiopathy in the first year of life. There is ventriculoarterial discordance resulting in parallel pulmonary and systemic circulations responsible for a state of hypoxia observed by central cyanosis. It is fatal unless mixing of oxygenated and deoxygenated blood occurs through patent ductus arteriosus (PDA), atrial or ventricular septal defect (ASD and VSD). The initial results of the Jatene procedure were investigated in infants with TGV.
Methods: A retrospective study from July 2008 to March 2015 analyzed 23 patients' medical records who underwent Jatene procedure at Esperança Hospital of Recife or PROCAPE. Exclusion criteria included age over 32 months, coarctation of the aorta, pulmonary stenosis or hypertension, and Taussig-bing syndrome. Age ranged from 4 days to 32 months (m = 101,2 days) and weight from 2,700 to 5,100 g (m = 3,400 g). Associated with TGV 12 patients had ASD and PDA (52,2%), 10 had VSD, ASD and PDA (43,5%) and only one had ASD and pulmonary artery banding (4,3%). The Jatene procedure is performed under general anesthesia with 8oC intermittent cardioplegia. The aorta and pulmonary artery are detached from their native roots, reattached to the opposing root, and the coronary arteries are transplanted from the aorta to the neo-aorta.
Results: Hospitalization period varied from 13 to 180 days (m = 37,1 days). Ten patients presented low cardiac output syndrome (43,4%), seven had renal failure with a need for peritoneal dialysis (30,4%), and seven were deceased (30,4%). Infants over 15 days had higher morbimortality rates as well as patients with ASD and VSD compared to those with ASD and PDA.
Conclusion: The Jatene procedure is a definitive treatment for TGV and has better results when done early, before 15 days of life.

P1334 - NURSE DRIVEN REMOVAL OF CHEST TUBE DRAIN

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Background: National Heart Institute nurse driven in removal of chest tube because all our patient post cardiac surgery has chest drain. Common chest drains that in place after cardiac surgery are such as pericardial, mediastinal or pleural chest tubes. Our main objective of this study is to present the best available evidence related to the Paediatric ward nurses in removal of chest drains. Secondary objective is to enable the registered nurse to remove chest drain with safety standard applied and developing their competency skill.
Materials/Method: This is a retrospective observational study to audit inpatient Paediatric General Cardiac ward nurses in removal

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of chest drain at Day 4 post-operative for patient above 1 year. The sample was collected from October 2013 to December 2014, which consist of 182 patients audited in removal of chest drain and total of 313 chest drains were removed. 243 chest drains was removed by the specialized cardiac nurses and another 70 chest drains was removed by surgeon. Audit form was used to collect the data. Training were conduct by Surgeon and Senior Nurse Mentor with workshop, bedside teaching and hand on. Inclusive and exclusive criteria were developed for removal of chest drain to prevent any complications and to ensure patient safety.
Results: From the data collected shows 243 chest drains were removed by ward nurses. 11 cases (3.7%) developed complication such as pneumothorax. 232 (95%) were successfully removed chest tubes without any complication.
Conclusion: Chest tube removal can be successfully performed by the specialized cardiac ward nurses with minimal complication. The advantage is reduce waiting time for surgeon for chest tube removal. Therefore it enhance early mobilization and recovery of Paediatric patient. Structured training in removal chest drain, assess and reassessment is very important element to be conduct in order to develop competent staff.

P1338 - LONG TERM OUTCOMES OF ISOLATED TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE REPAIR IN NEONATES AND INFANTS A 41 YEAR EXPERIENCE FROM A SINGLE INSTITUTION

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Background: Outcomes of surgery for total anomalous pulmonary venous drainage (TAPVD) has improved. However, surgery in the neonatal period and the development of postoperative pulmonary venous obstruction (PVO) is associated with high mortality.
Methods: A retrospective review was conducted for all neonates and infants (n = 214) undergoing surgery for isolated TAPVD (1973-2014). Multivariate analysis was performed.
Results: Preoperative PVO occurred in 112 (52%) patients. There were 91 (43%) supracardiac, 45 (21%) cardiac, 60 (28%) infracardiac and 18 (8%) mixed TAPVD. Median age was 18 days (1day-1 year). There were 17 (7.9%) early deaths. Risk factors for early mortality were prolonged cardiopulmonary bypass (CPB) time ($p = 0.005$) and neonatal age at surgery ($p = 0.048$). Era of surgery was not a risk factor. Early mortality for infants (n = 81) and neonates (n = 133) was 2.5% and 11%, respectively ($p = 0.021$). Hospital mortality according to era was 12% (6/49) (infant 9.1%, 1/11; neonates 13%, 5/38) during 1973-1988; 8.2% (5/61) (infants 4.8%, 1/21; neonates 10%, 4/40) during 1989-1998; and 5.7% (4/70) (infants 0%, 0/36; neonates 8.8%, 4/34) during 1999-2008 and 5.9% (2/34) (infants 0%, 0/13; neonates 10% 2, 2/21) during 2009-2014. There were 10 (5%) late deaths. Survival at 10 and 20 years was 88 ± 2.2% (95% CI:82-91%). Reoperation for postoperative PVO was required in 22 (10%) patients. Risk factors for reoperation were prolonged CPB time ($p = 0.015$), lower operative weight ($p = 0.003$) and an episode of postoperative pulmonary hypertensive crisis ($p = 0.005$). Freedom from reoperation at 10 and 20 years was 90 ± 2.2% (95% CI:85-93%) and 86 ± 3.2% (95% CI:78-91%), respectively. All survivors were asymptomatic at a mean of 13 ± 9 years (1 month-42 years) after surgery.
Conclusions: While isolated TAPVD repair in infants can be performed without mortality, it is associated with high mortality in neonates that remained unchanged over long study period. Survival beyond 1-year of age is associated with excellent long-term outcomes.

P1342 - INHALED NITRIC OXIDE WITH A NASAL CANNULA AND USE OF PHOSPHODIESTERASE INHIBITOR TO FACILITATE WEANING FROM INHALED NITRIC OXIDE FOR CHILDREN AFTER CONGENITAL CARDIAC SURGERY

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Objective: Inhaled nitric oxide is effective for critical pulmonary perfusion. After extubation, inhaled nitric oxide is usually discontinued and rebound pulmonary hypertension is the problem. The purpose of this study was to evaluate inhaled nitric oxide with a nasal cannula and use of phosphodiesterase inhibitor in facilitating its problem.
Methods: Inhaled nitric oxidewas administered in the case with critical pulmonary perfusion (central venous pressure >20 mmHg or pulmonary hypertensive crisis) during the operation or in the early postoperative period. In ten cases, inhaled nitric oxide could not be discontinued before extubation. In these cases, inhaled nitric oxide with a nasal cannula was continued after extubation and phosphodiesterase inhibitor was also used. Patients ranged in age from 2 months to 2.5 years and weighed from 2.5 kg to 12.7 kg. Four cases were after bidirectional Glenn anastomosis, three case were after total cavopulmonary connection and three cases were after patch closure of ventricular septal defect.
Results: There was no hospital death. No rebound pulmonary hypertension occurred in any cases. The time to discontinuation of inhaled nitric oxide after extubation raged from 2 days to 26 days. The dose of inhaled nitric oxide ranged from 1 to 10 ppm. Phosphodiesterase inhibitor (sildenafil in four cases or tadalafil in three cases) was administered before or after extubation. The dose of sildenafil ranged 0.5 mg/kg to 1.5 mg/kg and the dose of tadalafil was 1 mg/kg. No toxic side effect was observed in any cases.
Conclusion: Inhaled nitric oxide with a nasal cannula and use of phosphodiesterase inhibitor can facilitate weaning from inhaled nitric oxide for children after congenital cardiac surgery with critical pulmonary perfusion.

P1349 - ARTERIAL SWITCH OPERATION IN PATIENTS WITH TRANSPOSITION AND LEFT SIDED AORTA

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Background: The arterial switch operation (ASO) has evolved as the treatment of choice for infants with transposition. The procedure

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is technically challenging when the aorta is left-sided, and requires surgical expertise and precision for a good surgical outcome. We review here our experience with such patients.
Methods: We carried out the arterial switch procedure in 20 patients diagnosed as transposition with left-sided and anterior aorta, diagnosed between January, 2002, and October, 2013. Hospital records were analyzed for segmental anatomy, associated cardiac anomalies, the coronary arterial patterns, aortic cross clamp and cardiopulmonary bypass times, and specific surgical techniques used for transfer of the coronary arteries and reconstruction of great arteries. We recorded the outcomes in terms of in hospital survival and left ventricular function at most recent follow up.
Results: Age at the time of operation ranged from 3 days to 18 months (median- 75 days), and weight ranged from 3 kgs to 8.8 kgs (median -3.85 kgs).All patients survived the operation.We did not perform the LeCompte manouvrein 9 patients. Mean cardiopulmonary bypass time was 157.5 ± 24.9 minutes, (median 161 minutes); mean aortic cross-clamp time was 101.2 ± 23.8 minutes, (median 102 minutes). One patient died on the second postoperative day due to sudden onset of ventricular fibrillation. Follow-up ranged from 23 to 31 months(mean 28.04 ± 2.32 months, median 28.4 months). At last follow-up, all 19 survivors are in NYHA class I, and none require cardiac medications. Serial echocardiograms have revealed normal biventricular function and no residual defects.
Conclusion: With appropriate technical modifications, patients with concordant atrioventricular and discordant ventriculo-arterial connections with left-sided aorta can undergo successful anatomical repair.

P1350 - A SINGLE STAGE SURGICAL REPAIR OF INTERRUPTED AORTIC ARCH PROXIMAL AORTO PULMONARY WINDOW TYPE 1 WITH AORTIC ORIGIN OF THE RIGHT PULMONARY ARTERY

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Introduction: The combined deformation consisting of an interrupted Aortic Arch, a proximal aorto-pulmonary window type 1, aortic origin of the right pulmonary artery, intact ventricular septum and patent ductus is a rare and surgically challenging disorder. We present single-stage-repair and successful perioperative management of such a patient.
Case Description: The patient was born in 41 + 1 week of pregnancy with bodyweight of 3270 gram in a district hospital. Following pathological pulse-oxymetric screening echocardiography was performed. After initial diagnosis of isthmus-stenosis and an aortopulmonary window, the 2-day-old neonate was referred to our ICU, presenting with tachypnoea of 70/min without other signs of congestive heart failure or hereditary malformations. Another echocardiography revealed the interrupted aortic arch, a proximal aorto-pulmonary window type 1 and under prostaglandin-therapy still patent ductus. Hence the single-stage-repair was performed. After harvesting the pericardial patch, the ductus was ligated and the aortic stenosis dissected. Following the preparation and attachment of the descending aorta to the aortic arch, the patch was sutured to the anterior half of the proximal descending aorta. Then the suture line descended to the posterior inner wall of the ascending aorta, leaving the abnormally positioned pulmonary artery on the left side of the patch. The remaining part of the aortic wall was now attached to the patch and the pulmonary artery on the other side as well. Postoperative care was uneventful.
Discussion: This rare and complex malformation requires fast and sufficient diagnosis and therapy. Due to critical condition of the mostly just days old patients, vast and invasive diagnostics are irrelevant. Echocardiography is, if performed by experienced physicians, the diagnostic method of choice. Because of low impact on the therapeutical approach angiography is mostly obsolete. A fast and complete surgical repair is essential to prevent pulmonary damage and to restore perfusion in the lower body.

P1352 - GIANT PSEUDOANEURYSM OF THE RIGHT VENTRICULAR OUTFLOW TRACT AFTER SURGICAL COMISUROTOMY OF PULMONARY VALVE SURGICAL APPROACH

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Background: Ventricular pseudoaneurysm result from rupture of myocardium with resultant localized hemopericardium. Right ventricular pseudoaneurysms are exceptionally rare, and can be a complication of cardiac surgical procedures done for congenital heart disease1 2. This can have catastrophic consequences3; so, early repair is of vital importance. We describe a case in a boy who developed a giant pseudoaneurysm three years after surgical comisurotomy of pulmonary valve for severe stenosis and RVOT ampliation with bovine pericardial parch and the surgical repair.
Case Report: A 15-years-old boy with Noonan syndrome, and diagnosed (at the age of 12 years) with Severe Pulmonary valve stenosis, was attended prior surgery at age 12 years: Through right ventriculotomy, Pulmonary Valve comisurotomy and right ventricular outflow tract ampliation with bovine pericardiac patch was achieved. Presented immediate bleeding, needing reoperation, without findings of specific point of bleeding, he was stayed in ICU during 2 days, total In-hospital stay of 10 days. At 3 years postoperative follow up the patient was consulting having dyspnea and precordial pain during ordinary physical activity. During examination he was found to have a pseudoaneurysm of right ventricular outflow tract (RVOT) by transthoracic echocardiogram (TTE). The patient underwent elective surgery, arterial and venous femoral cannulation, establish cardiopulmonary bypass, careful sternotomy was carried out, pseudoaneurysm was dissected, and under the beating heart on full cardiopulmonary bypass, the pseudoaneurysm was opened and the communication with the RVOT was confirmed and closed with a bovine pericardial patch using a continuous suture technique, reinforced with the pseudoaneurysm wall in a layer over layer manner, without postoperative complications.
Conclusions: Opportune repair of ventricular pseudoaneurysm is of vital importance4. Echocardiography is an excellent diagnostic method for this entite3 5. Althought midline sternotomy could be safe in most patients; peripheral cannulation should be considered when the pseudoaneurism adheres to the sternum.

P1353 - IMPROVING REALISM DURING PEDIATRIC ECMO SIMULATION

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Background: According to the Extracorporeal Life Support Organization (ELSO) guidelines, "low volume" Extracorporeal Membrane Oxygenation (ECMO) programs (<20 cases/year) may need additional continuing education for all team members. Simulation based training is increasingly recognised as a powerful educational tool to improve safety, attitudes and teamworking surrounding emergencies with pediatric ECMO.
Methods: In order to increase the fidelity of our paediatric ECMO emergency scenarios, we established the ability to simulate the different colours of oxygenated and deoxygenated blood of the ECMO circuit, allowing us to reproduce gas supply failure and oxygenator failure scenarios. Porcine blood was used to prime our modified paediatric ECMO circuit. Our standard circuit consists of a centrifugal pump and a pediatric heparin coated hollow fiber oxygenator. We modified it to include a "de"-oxygenator and a reservoir bag inserted in series and hidden from view. This spare oxygenator is used to deoxygenate the blood by running a sweep gas of 100% carbon dioxide. The system is also used to simulate air entrainment into the circuit. The presence of blood demonstrates how easily it foams, making the scenario more realistic and difficult to deal with.
Results: This technique has been used 8 times, training 160 team members since 2013. An easily discernible difference in line colour has occurred each time. Participants strongly noted the ease of differentiating oxygenated and deoxygenated blood. Feedback supports the realism of the technique compared to clear prime simulation. Detailed analysis of the feedback will be presented.
Conclusions: Use of porcine blood in the ECMO circuit with a "de" oxygenator is feasible and improves the fidelity of the simulation training experience. Allowing oxygenated and deoxygenated blood to be recognised permits realistic troubleshooting and problem solving in ECMO emergencies such as gas supply failure, oxygenator failure and air entrainment.

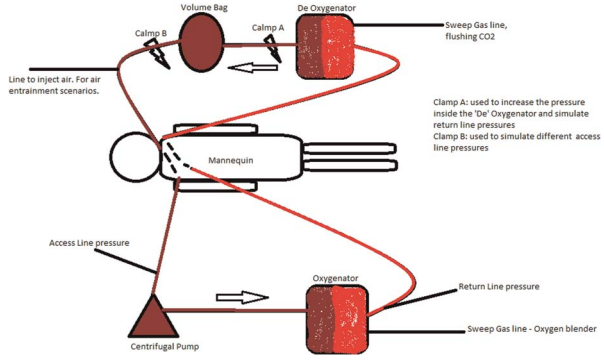

Figure.

P1367 - IMPROVED PROGNOSIS IN A TWO STAGE APPROACH FOR SURGICAL CORRECTION OF VENTRICULAR SEPTAL DEFECT IN PATIENTS WITH TRISOMY 18

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Background: A congenital heart defect, particularly ventricular septal defect (VSD), is common in patients with trisomy 18 (T18). The indication for cardiac surgery in T18 patients is still controversial and requires discussion from the perspectives of postoperative outcomes and prognosis, in addition to bioethics.
Patients and Methods: The prognosis of surgical correction of VSD with T18 was determined in a retrospective review of medical records of 30 patients with T18 who were followed in North Osaka territorial hospitals in Japan from April 2008 to May 2016.
Results: Of the 30 patients, 16 (53%) underwent surgical treatment after careful informed consent was obtained from their parents (n = 15, VSD; n = 1, coarctation of aorta with VSD). One (6%) of these 16 patients underwent primary correction at 303 days after birth (BW: 3.5 kg) and 15 (93%) underwent first-stage palliative surgery (n = 14, pulmonary artery banding (PAB); n = 1, bilateral PAB, all with PDA ligation) from 12 to 126 days after birth (median: 127 days) (BW: 1.0 to 2.8 kg, median: 2.2 kg). In these 15 patients, there were three deaths (20%) due to pneumonia (n = 2) and sudden death (n = 1). Eleven (73%) of the 15 patients underwent second-stage correction (VSD closure and PAB debanding) 1.5 to 14.8 months (median: 8.8 months) after PAB (BW: 2.1 to 6.8 kg, median: 4.8 kg) and one is waiting for second-stage correction. Survival ranged from 15 to 85 months (median: 36.5) after second-stage correction and patients were discharged with improved symptoms. Nine (75%) of 12 patients underwent tracheostomy before correction. These 12 patients did not have severe gastrointestinal anomalies such as esophageal atresia.
Conclusion: A two-stage approach for surgical correction of VSD was practical and effective in carefully selected patients with T18. Long-term follow up is necessary to confirm the efficacy.

P1370 - THE FATE OF THE TRANSLOCATED AORTIC ROOT AFTER THE MODIFIED NIKAIDOH OPERATION

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Background: The modified Nikaidoh operation in transposed great aretries, ventricular septal defect and pulmonary stenosis allows translocation of the whole aortic root along with both coronary arteries.
Aim: We retrospectively analyzed our results with emphasis on the translocated aortic root dimensions.
Methods: Inbetween july 2012 and july 2016 7 patients underwent the modified Nikaidoh procedure.Four patients had DTGA/VSD/PS and 3 had DORV/VSD/PS anatomy. In all patients the aortic root was excised and translocated with attached both coronaries. The VSD was closed by a dacron patch in 5 and directly in 2 patients.The pulmonary artery was directly connected to the RVOT after a LeCompte manuever.
Results: Median age at operation was 1 year(30 days-3 years), median weight 9.4 kg (3.2-11 kg) and median follow up 3.6 years (2.7- 4 years).Median duration of extracorporeal circulation and aortic crossclamp were 190 min (150-347 min) and 90 min (43-108 min) respectively. One patient died 47 days postoperatively due to multiorgan failure following cardiac transplantation after unsuccsessfull attempts of closure of multiple muscular VSD´s. At the follow up there was no gradient on the LVOT in any patient and mild aortic regurgitation in one patient. The aortic annulus Z score decreased sligtly from 3.4 (2-4.3) preoperatively to 2.7 (1.7–4) at the follow up (p = 0.08).The aortic root Z score increased insignificantly from 1.2 (0.4-4) preoperatively to 2.1 (0.1-4.4) at the follow up (p = 0.66).The ascending aorta Z score

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remained stable from 0.9(0.4-4)preoperatively to 0.5 (0-3.5)at the follow up (p = 0.35).
Conclusions: The modified Nikaidoh operation offers unobstructive pathway from the left ventricle and preservs the aortic valve function. The Z scores of aortic dimensions indicate, that the aortic annulus, root and ascending aorta remain stable in the midterm.Further follow up is necessary to asess the longterm function of the translocated aortic valve and the root.

P1375 - RESPIRATORY VIRUSES STILL A MAJOR PROBLEM IN PEDIATRIC CARDIAC SURGERY PATIENTS

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Background/Objective: Respiratory viral infections (RVIs) are critical events in infants and children with congenital heart diseases, particularly when they occur during hospital stay for cardiac surgery. However, they are relatively rare events. We therefore used sophisticated statistical armamentarium (generalized linear models – GzLMs) to analyse risk factors and consequences of RVIs in this population.
Patients/Methods: Of 1797 patients admitted from January 2012 to December 2014, 209 pediatric cardiac surgery subjects (age 1-14 months, median 7 mos.) had clinical suspicion of RVIs during hospitalization, with nasopharyngeal aspirates available for analysis. Immunofluorescence assays were performed for detection of influenza and parainfluenza viruses, adenovirus and respiratory syncytial virus (RSV). The periods of observation were registered for construction of GzLMs.
Results: Viruses were detected in 22% of instances; RSV was the most prevalent one (10%) followed by parainfluenza (9%). We used Poisson regression to demonstrate a higher rate of infections in 2013 relative to 2012 and 2014 (p = 0.040). Weight quartiles 2 and 3 (lowest to highest) were associated with risk (p = 0.051 and p = 0.039 respectively) compared to quartile 1 (relative protection). We used ordinal regression to analyse the impact of age, weight, Down syndrome, type of cardiac anomaly and RVIs on the length of hospital stay and occurrence of fatal events. Weight appeared as a protective factor (for quartiles, HR, 0.79; 95% CI, 0.71-0.88); AV canal defects/left isomerism and cyanotic defects with right heart obstruction were "high risk" compared to simple shunts (respective HR with 95% CI, 3.16[1.27-7.84] and 2.21 [1.11-4.39]). RVIs were associated with prolonged hospitalization (HR, 2.11; 95% CI, 1.16-3.83), with an 18-day increase in hospital stay (p = 0.002).
Conclusions: In addition to the known seasonal variations, there are year-to-year changes in the risk of RVIs in hospitalized patients. Low weight, complex and right-heart obstructive anomalies, and RVIs are important determinants of prolonged hospital stay.

P1379 - AUTOIMMUNE VASCULITIS IN A CHILD FOLLOWING TETRALOGY OF FALLOT REPAIR IN UGANDA

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Background: Tetralogy of Fallot is one of the conotruncal abnormalities associated with 22.q.11 deletion syndrome which presents with thymic hypoplasia, hypocalcaemia and have an increased risk of autoimmune disorders. We report about a child post TOF repair who presented to the Uganda Heart Institute diagnosed to have autoimmune vasculitis.
Clinical Case: An 11 year girl who had undergone TOF repair 8 years prior to the illness presented with persistent high grade fevers for 2 weeks, abdominal pain, petichae with no arthralgia. Clinical examination revealed a sick child febrile 39 °C, with petichae on palms and sole of the feet. Oral thrush and bleeds in the mouth and no dysmorphism. She also had blisters on the lower limbs with secondary infection. She was hypoxic with oxygen saturation of 86% on room air. Blood pressure 88/56 mmHg with grade 3/6 ejection systolic murmur in the left upper sternal edge. Baseline laboratory results upon admission showed leukocytosis, WBC 17.9 × 103, with a neutrophilia, 16 × 103(89%), Haemoglobin 12.3 g/dl, thrombocytopenia 93 × 103 /µl and elevated C- reactive protein of 80 mg/l(NR <6 mg/l). The peripheral film revealed macro platelets, left shift and no schistocytes. The electrolytes were deranged; hypocalcaemia 1.6 (2.1-2.5), hypokalemia 2.6 mmol/L and hypomagnesaemia 0.6 mEq/L. Thyroid function tests and PT/APTT were normal. No organisms were grown on blood culture. urine analysis revealed yeast cells and cultured >105 candida albicans. Despite antibiotics, she worsened with headache, seizures and loss of consciousness. Cardiac echo showed intact VSD patch, moderate PA conduit stenosis and no vegetations. Brain CT was normal. Autoimmune panel C-ANCA was elevated (18U/ml). Intravenous methyl prednisolone was initiated and the child regained consciousness in 24 hours.
Conclusion: Tetralogy of Fallot patients may manifest with hypocalcaemia, severe infections and have a tendency of autoimmune disorders. A multidisciplinary model is necessary in the management of these patients.

P1384 - CARDIAC ECMO IN A MEDIUM-SIZED CONGENITAL HEART UNIT: THE CASE FOR INCREASING AVAILABILITY WORLDWIDE

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Background/Hypothesis: Survival free of major complications is uncertain in children on VA-ECMO for cardiac failure. Current literature maintains volume is the key quality indicator for cardiac programs.
Materials and Methods: We reviewed outcomes of all children on VA-ECMO in a medium-sized congenital heart unit over a 4-year period (June 2012-December 2016). Primary outcomes were survival to de-cannulation and discharge, and current survival status. Secondary outcomes included bleeding events, cannulation site infection, blood/blood product utilization, degree of cardiac recovery, return to catheterization lab or operation room (OR), days on VA-ECMO and current functional status.
Results: 29 children were supported on VA-ECMO: 24 post cardiac surgery; 1 cardiac failure; 2 septic shock; and 2 intractable

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arrhythmias. 12 were placed on VA-ECMO post single-ventricle repair; 10 from cardiac arrest; and 5 from weaning failure from CPB. Median length of ECMO was 5 days (6 hours-18 days). 11 required re-exploration for bleeding and 7 required return to the OR and/or catheterization lab. 27 (93%) were de-cannulated successfully; and 23 (79%) were discharged home. There have been no late deaths. 2 remain with tracheostomy; 1 required femoral artery reconstruction; 1 required stent placement in the SVC secondary to cannulation for 18 days; and 1 patient had loss of digits secondary to DIC and small vessel thrombosis. Post-operative ECMO represented 2.2% of major cardiac cases during this period.
Conclusions: Excellent outcomes can be achieved in very high-risk VA-ECMO candidates in medium-sized cardiac units. A professional circuit team, intensive cardiac functional monitoring and daily multidisciplinary rounds are critical for optimal outcomes.

P1388 - OUTCOMES OF TRIFECTA VALVE IN THE PULMONARY POSITION: A SINGLE-CENTER EXPERIENCE

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Objective: Evaluate early and midterm outcome of pulmonary valve (PV) replacement using Trifecta (St. Jude Medical, Inc.) bioprosthetic valve.
Methods: 28 patients underwent PV replacement with Trifecta valve (October 2012-December 2014). Median age was 12.0 years (3.2-62.6 years). Primary diagnosis was Tetralogy of Fallot (n = 13, 46.4%). Preoperative diagnosis was PV insufficiency (n = 23, 82.1%). Redo sternotomy was performed in 25 patients (89.3%). Median implanted valve size was 25 mm (19mm-29mm). Median length of stay was 5 days (3-53 days). Median follow-up was 14.0 months (6.6-26.3 months). Echocardiograms were evaluated by 3 cardiologists and scored for pulmonary insufficiency, right ventricular (RV) size, and PV gradient. Echocardiograms were designated early post-operative (median 22.3 days), intermediate post-operative (median 119 days), and late post-operative (median 425.9 days). Numerical values were assigned: normal = 1, mild = 2, moderate = 3, and severe = 4 for pulmonary insufficiency and RV size. Mean scores were obtained to determine final classification.
Results: There were no early or intermediate valve-related deaths. In-hospital mortality was nil (n = 0). No patients required replacement of PV prosthesis within the study period. No post-operative valve thrombosis was noted. Early echocardiography results showed a decrease in pulmonary insufficiency and RV size with mean scores of 2.3 and 1.4 respectively. At last follow-up, 3 patients (14.2%) were found to have moderate to severe PV insufficiency (score ≥3). Peak PV gradient was 12.5 + /-5.6 at last follow-up. RV size was normal or mildly dilated in 24 (85.7%) patients at last follow-up compared to 24 (85.7%) patients with severely dilated ventricles preoperatively.
Conclusions: Trifecta valve can be placed in the pulmonary position with good early results; care must be given to the position of the valve to ensure leaflets are not impinged upon by surrounding native pulmonary artery tissue.

P1412 - CUSTODIOL VERSUS CONVENTIONAL CARDIOPLEGIA IN TETRALOGY OF FALLOT SURGERY

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Background: We examine whether Custodiol and conventional cardioplegia have differential effects on patients' perioperative and postoperative conditions.
Materials and Methods: The initial sample consists of 90 pediatric patients with Tetralogy of Fallot (TOF) who underwent a total correction surgery from March 2014 to November 2016. The patients were randomly allocated to receive Custodiol or conventional cardioplegia. Both groups of patients' perioperative and postoperative conditions were collected. We employed ordinary least square (OLS) regressions and probit models to test our hypotheses. Perioperative conditions include cardiopulmonary bypass (CPB) and aortic cross-clamping (ACC) time. Post-operative conditions consist of right and left ventricular ejection fraction (RVEF and LVEF), mechanical ventilation time, ICU stays, overall stays, arrhythmia, complications and Vasoactive Inotropic Score (VIS).
Results: Based on the final sample of 78 patients, we find that Custodiol and conventional cardioplegia have the differential effects on CPB time, ACC time and post-operative outcome for patients undergoing TOF surgery. The results indicate that patients who received conventional cardioplegia tend to have shorter CPB time than those receiving Custodiol. In addition, patients who received conventional cardioplegia tend to have shorter ACC time than those receiving Custodiol. These results remain evident after controlling for operation procedure (i.e. transannular patch vs. nontransannular patch), other lesions (e.g., PDA, ASD), and the presence of previous shunt. In addition, there is evidence to suggest that CPB time had a positive effect on VIS during the first 24 hours after surgery and had a negative effect on RVEF. Overall, there is no evidence to support the notion that CPB time had any effect on postoperative mechanical ventilation time, ICU stays, overall stays, arrhythmia, and complications.
Conclusions: Relative to the use of conventional cardioplegia, the use of Custodiol results in longer CPB time, longer ACC time and higher VIS during the first 24 hours after surgery.

P1413 - 39 YEARS OF EXPERIENCE OF TREATING TRANSPOSITION OF THE GREAT ARTERIES IN A SINGLE LOW VOLUME CENTER

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Introduction: The success rate of arterial switch operation (ASO) depends on the experience of the congenital heart surgery team, which in turn depends on the case volume of the surgical center.
Objective: We report our 39 year experience in ASO's and follow-up results.
Methodology/Statistical Analysis: Between January 1977 and December 2016, 115 patients underwent ASO at our center. The patients were divided into three groups according to the time period in which they were operated. Group-1 (n = 18) was

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operated from 1977 to 1998. Group-2 (n = 24) was operated during the international knowledge transfer program between Wessex Cardiothoracic Center, Southampton General Hospital and our center from 1999 to 2002. Group-3 (n = 73) was operated in post-program years (2003-2016). Patients were followed for early and late mortality, postoperative morbidity and reintervention or reoperation.
Results: Median follow-up time was 5.1 years (Interquartile range (IQR) 2.76-9.1 years). A median number of 2 (IQR 0-6) procedures/year is performed at our center. Early mortality in groups was 77.8% vs 41.6% vs 4.1% accordingly ($p < 0.001$). The Kaplan-Meier estimated 15-year-survival-rate in groups is 6.3%, 44.5%, 91.4% respectively ($p < 0.001$). In total 6 (5.2%) patients died during the late postoperative period and 7 (6.1%) were lost to follow-up. 25 (21.7%) of the followed patients have mild to moderate aortic valve insufficiency. 4 (3.5%) patients underwent a balloon angioplasty for re-coarctation of the aorta, and 4 (3.5%) for pulmonary artery stenosis. 1 (0.86%) patient underwent right ventricle infundibulectomy and bidirectional Glenn shunt. 2 (1.7%) patients have severe development impairment.
Conclusions: Knowledge transfer programs between high expertise pediatric cardiac surgery centers and low-volume centers can be beneficial in lowering early post-operative mortality rates. It is possible to perform arterial switch operations safely and with good outcome in a low-volume center.

P1422 - AORTIC VALVE REPAIR WITH PARTIAL LEAFLET SPARING METHOD IS THERE ANY POTENTIAL TO GROW

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Objective: Aortic valve repair has encouraging midterm results in selected patients. However, in terms of the growth of aortic valve, there has not been reported. We introduced aortic valve plasty (AVP) with partial leaflet sparing method. We try to preserve leaflet tissue as much as possible.
Methods: Between September 2004 and July 2016, eleven pediatric patients with aortic valve regurgitation (AR) underwent primary aortic valve repair with partial leaflet sparing electively. A variety of techniques were used in performing partial leaflet sparing AVP, depending on the specific cause of regurgitation in each patient.
Results: The age (median) and body weight at the operation were 5.4 years (1.7–16) and 14.0 kg (9.1–44.6). There was no mortality. The follow up was 100%. The follow up period (median) was 5.6 (0.4–11.2) years. The previous operations prior to AVP were performed in 6 patients (VSD closure (n = 2), AVSD repair (n = 1), Norwood (n = 1), arterial switch operation (n = 1), PDA division (n = 1)). And concomitant procedures are following: VSD closure (n = 4), mitral valve repair (n = 2), mitral valve replacement (n = 1), RVOTR (n = 1) and TCPC (n = 1). There were bicuspid aortic valve in 1 patient and tricuspid aortic valve in 10. The one leaflet extension was performed in 6 patients, one cusp replacement with using pericardium in 1, patch augmentation in 2, and leaflet plication in 2. Leaflet slicing was performed, if needed. All pericardium was treated with glutaraldehyde. The recent echo showed that AR ($p = 0.0011$) and LVDd ($p = 0.019$) were improved significantly, compared with pre-OP and the aortic valve size was growing within normal range (Table). Freedom from reoperation was 100% at 10 years.
Conclusions: The present method with partial sparing of aortic leaflet can yield the growth potential of aortic annulus to pediatric patients with AR as well as effective reduction of the degree of AR..

Table 1. Results form recent Echocardiogram

	Pre-valve repair	Recent echo	P value
AR	3.1 ± 0.4	1.8 ± 0.7	0.0011
LVDd (% of normal)	126.2 ± 13.9	102.9 ± 24.6	0.019
Aortic valve diameter (% of normal)	125 ± 40	142 ± 37	0.41

AR: aortic regurgitation, Scoring of AR grade (1 in trivial, 2 in mild, 3 in moderate, 4 in severe); LVDd: left ventricular diastolic dimension

P1425 - INCREASED PRESSURE IN A PULMONARY ARTERY IS IT A CONTRAINDICATION TO BIDIRECTIONAL CAVOPULMONARY CONNECTION

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Background: Bidirectional cavopulmonary connection (BCPC) is optimal intermediate stage for hemodynamic correction of congenital heart diseases with single ventricle. There is no identical opinion upon optimal criteria for BCPC performance.
Materials and Methods: Over the 2012 to 2014 period 89 patients with SV at the age from 2 months to 13 years old (Me = 0,64; IQR:0,5-1,18) were involved into the study. They were performed catheterization of heart cavities with pulmonary resistance calculation at different stages of hemodynamic correction. The patients were divided into two groups: the first group (n = 64) – children with initial pulmonary artery (PA) pressure under 15 mm Hg, the second group (n = 25) – patients with PA pressure above 15 mm Hg.
Results: The average PA pressure was 11 mm Hg (IQR:10,00–13,75), indexed pulmonary blood flow volume (iQp) - 3,57 (mL/Min)/M2 (IQR; 2,69-4,87), indexed pulmonary resistance (iRp) - 1,41 Ед.Wood/м2 (IQR;0,90-1,98) in the first group. In the second group the average PA pressure was 20 mm Hg (IQR:17,00-26,00), iQp 4,56 (mL/Min)/M2 (IQR: 3,17-6,04), iRp 2,36 Ед.Wood/м2 (IQR:1,85-3,10). Taking into consideration increased PA pressure, oxygen test was performed. A slight decrease of average PA pressure up to 19 mm Hg (IQR:17,00-30,00),), $p = 0,02$, iQp increase up to 11,2 (mL/Min)/M2 (IQR:5,85-15,6), $p = 0.003$) and iRp decrease up to 1,37 un. Wood/м2 (IQR:0,72-1,95), $p = 0,002$ were marked after the test. All the patients were performed BCPC. Before the total cavopulmonary connection the average PA pressure was 8 mm Hg (IQR:7,00–9,13), iQp 2,69 (mL/Min)/M2 (IQR;2,14-3,15), iRp was 1,04 un.Wood/м2 (IQR;0,81-1,57), $p = 0,1$ in the first group. In the second group it was 8 mm Hg (IQR:6,38–11,25), iQp 2,46 (mL/Min)/M2 (IQR;2,22-3,30), iRp was 1,23 un. Wood/м2 (IQR:0,86-2,3). Significant difference between two groups was not disclosed, $p > 0,05$.
Conclusion: Thus, the pressure in a pulmonary artery higher than 15 mm Hg is not a contraindication to BCPC.

P1426 - HAND SEWN VALVED BOVINE PERICARDIAL CONDUITS FOR RIGHT VENTRICULAR OUTFLOW TRACT RECONSTRUCTION A SINGLE CENTER EXPERIENCE

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Background: Right ventricle to pulmonary artery conduit is an essential part of treatment in some forms of congenital heart disease. Ideal conduit should be easily available, cost effective and should have longevity. There are various conduits available but they do not demonstrate growth potential, which creates patient prosthesis mismatch as the child grows. There are some reports that decellularised bovine pericardium may have growth potential due to repopulation by the host cells. This study reviews our experience with implantation of hand sewn valved bovine pericardial conduits for right ventricular outflow tract reconstruction.
Materials and Methods: 18 patients underwent right ventricle to pulmonary artery reconstruction using hand sewn decellularized bovine pericardial conduits; material used for constructing the valve being either decellularized bovine pericardium or 0.1 mm polytetrafluoroethelene. We analyzed the preoperative characteristics, intra-operative details and short term results including mortality.
Results: Mean age of the patients was 45.9 ± 74.6 (Range: 1-312) months. Mean weight was 11.6 ± 11.2 (Range: 3.7-51.2) kilograms and height 80.3 ± 27.6 (Range: 54-167) centimeters. Mean body surface area was 0.49 ± 0.31 (Range: 0.23-1.54) M2. Mean expected pulmonary artery size with respect to body surface area preoperatively was 11.4 ± 2.3 mm (Range: 8.4-17.6). Mean implanted conduit size was 14.1 ± 2.0 mm (Range: 12-18). Mean CPB time was 235 ± 62 (Range: 88-360) minutes and aortic cross clamp time was 125 ± 45 (Range: 44-229) minutes. There was no mortality in the immediate postoperative period or follow up. Four out of eighteen (22%) patients had mild valvar regurgitation in the follow up echo. One patient out of eighteen (5.5%) required pulmonary artery stenting in the post operative period.
Conclusion: Hand sewn bovine pericardial conduits used for right ventricular outflow tract reconstruction provide effective palliation with excellent short term conduit function. Continued long term follow up studies are necessary to determine the perceived growth potential of the decellularized tissue.

P1427 - SERIES OF DIRECT CLOSURE OF TYPE 1 AORTOPULMONARY WINDOW WITH GOOD SHORT AND MID TERM RESULTS; SAFE AND COST EFFECTVIE TECHNIQUE

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Background: We are reporting here case series of sixteen cases of APW during the last five years out of which ten patients were done through direct surgical closure. Direct closure has advantage of avoiding cardio pulmonary bypass (CPB) and post op sequelae of CPB.
Materials and Methods: From January 2010 to December 2015, sixteen infants with Aortopulmonary window were operated in the department of Paed.cardiac surgery, Institute of children health Lahore, Pakistan. Out of Sixteen, ten were type 1 APW, in whom Operative technique included direct closure of the defect without using CPB. Operative technique included careful dissection, going around and getting control of APW, observing the ECG changes and confirming the absence of distortion of right pulmonary artery (RPA) and residual flow through echo. Immediate concern of direct ligation of APW is distortion of aortic valve, pulmonary valve, left coronary ischemia and obstruction of origin of right pulmonary artery. These ten patients were further analyzed for operative technique and clinical outcome. Age at operation ranged from 1.5 to 10 months (median 5 months). Weight at operation ranged from 3 to 6 kg (median 5 kg).
Results: Follow up ranged from 2 months to 48 months. There were no early or late deaths postoperatively. There were no residual defects or distortion of RPA in our series. ICU stay ranged from 1 to 2 days postoperatively.
Conclusion: Direct closure of type 1 APW is safe and has excellent short and mid term results in terms of cost and health of patient especially in those countries where limited resources are main factors affecting management of patients.

P1439 - AGGRESSIVE RECONSTRUCTION OF PULMONARY ARTERIES AT THE TIME OF SYSTEMIC TO PULMONARY SHUNT PLACEMENT IN NEONATES REDUCES THE RISK OF REINTERVENTIONS

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Objectives: the purpose of present study was to evaluate whether pulmonary artery patch repair at the time of shunt placement is beneficial in terms of reintervention rate.
Methods: 27 neonates with pulmonary atresia and confluent pulmonary arteries were included in the study. Following methods were used to create shunts: modified Blalock-Taussig shunt (n = 14, 52%), side-to-side aorto-Gore-Tex central shunt (n = 13, 48%). In 9 cases (group 1) where ductal insertion distally from the ostium of either left or right pulmonary artery was noted, relevant pulmonary artery was repaired with autopericardial or pulmonary homograft patch. In other 18 cases (group 2) only shunt placement was performed. Decision to perform pulmonary artery repair was based on surgeon's judgment. Surgical mortality and pulmonary arteries obstruction, required re-intervention prior to next stage or repair were recorded. Chi-square Pearson's test and Fisher's exact test were utilized to compare outcomes between groups.
Results: In group 1 there we no pulmonary artery obstructions, requred reintervention. In group 2 there were 8 cases (44%) of pulmonary artery obstruction, required reintervention. p = 0.015.
Conclusions: Pulmonary artery reconstruction at the time of systemic-to-pulmonary shunt placement in neonates with pulmonary atresia reduces the risk of re-interventions for pulmonary artery obstructions. Aggressive patch repair may be beneficial even in cases of usual ductal incertion.

P1440 - SHORTER CROSS CLAMP TIME AND SELECTIVE CORONARY PERFUSION IMPROVE SURVIVAL AFTER STAGE I PROCEDURE

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Objectives: The purpose of present study was to assess different myocardial protection strategies and their impact on stage-I outcomes.
Methods: 59 newborns underwent Stage-I procedure during 2005-2015 were include in the study. 28 patients (group 1) were operated on moderate hypothermia (26 C) and two-regional perfusion

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(selective antegrade cerebral and coronary) during distal arch reconstruction, followed by cordioplegic cardiac arrest and selective cerebral perfusion during proximal arch and accending aorta reconstruction. 31 patients (group 2) were operated on deep hypothermic circulatory arrest (18 C) with and without selective antegrade cerebral perfusion during all stages of aortic reconstruction. There were no significant difference between groups by BSA, urine output and serum lactate level prior to surgery (p is 0,311; 0,409 and 0,206 respectively). Mortality rate was compared between groups.
Results: Cross-clamp time (mean): group 1 - 63,1 ± 5,8 min., group 2 - 87,2 ± 3,1 min. (p = 0.001). 30-days mortality: group 1 - 25%, group 2 - 53% (p = 0.028).
Conclusions: Shorter aortic cross-clamp time achieved by utilizing two-regional perfusion improves myocardial function and reduces 30-days mortality rate after Stage-I procedures.

P1441 - EFFECTS OF POSTURAL CHANGE ON OXYGEN SATURATION IN PATIENTS AFTER THE FONTAN OPERATION COMPARISON OF FENESTRATED AND NON FENESTRATED PATIENTS

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Purpose: The aim of this study was to compare oxygen saturation alteration between fenestrated and non-fenestrated Fontan patients.
Method: We divided 26 patients into 2 groups: 7 patients who had fenestrated Fontan (group F) and 19 patients who had non-fenestrated Fontan (group NF). SpO2 were measured by pulse oximeter in the supine and sitting positions. In F (3male, 2-9year-old: median 5), all 7 patients were performed Extracardiac TCPC. In NF (9male, 3-31year-old: median 7), 3 were APC, 13 were Extracardiac TCPC, and 3 were Extracardiac TCPC with naturally closed fenestration.
Results: In NF, SpO2 changed from 92-98(mean; 95)% to 91-97 (mean; 94)% (p < 0.01), which was the same result of previously published report. Contrary, in F, SpO2 changed from 81-91 (mean; 89)% to 84-94 (mean; 91)% (p < 0.01).
Considerations: The blood flow via fenestration is much more due to IVC's flow. In sitting, the fenestration flow might reduce than that in the supine. First, in sitting position, to keep adequate brain blood flow, SVC flow should increase. Second, IVC flow will decrease because resistance of lower limbs:LL will become bigger, or because much more blood will be kept in LL during sitting.
Conclusion: There are some study limitations. First the number of patients was still small. Second, changes of blood flow through fenestration were'nt checked. But we demonstrated in Fontan patients with fenestration SpO2 was a little bit increase during sitting than supine position. We think SpO2 itself should increase when fenestration naturally closed, but the patterns of SpO2 alteration with postural change will suggest an occlusion of the fenestration.

P1442 - NATIVE AORTIC COARCTATION IN NEONATES AND INFANTS COMPARISON OF OUTCOMES WITH BALLOON COARCTOPLASTY AND SURGERY

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In neonates and infants with severe native aortic coarctation, surgical repair is the standard therapy while balloon angioplasty remains controversial. At our institution, balloon coarctoplasty was initially restricted to critically ill patients as a salvage treatment. However, with good immediate results in these patients, we sought to compare our midterm outcomes of balloon and surgical coarctoplasty in infants.
Methods: Retrospective records review: we analyzed all patients with coarctation who underwent balloon or surgical coarctoplasty at our center with age <1 year at presentation and >6-months post-procedure follow up. Patients with significant aortic arch hypoplasia, long segment coarctation and associated intracardiac lesions were excluded. Clinical, echocardiographic and procedural details were recorded at initial presentation and follow-up. Need for reintervention was noted.
Results: 75 total patients: 28 balloon coarctoplasty (group A), 47 surgical coarctoplasty (group B). Table 1. There were two deaths (immediate mortality 4.2%) in group B, and none in group A. For neonatal presentation, group A patients had a significantly higher reintervention rate than group B (p = 0.006). On logistic regression analysis, balloon coarctoplasty and residual coarctation post-procedure were significant predictors of reintervention. On Kaplan-Meier analysis with neonatal presentation, freedom from reintervention in group B was significantly higher than group A (P = 0.0036). Freedom from reintervention was also greater for group B as a whole, and conversely greater for group A than group B for presentation between 1-12 months (not statistically significant).
Conclusion: Our results demonstrate balloon coarctoplasty is a viable alternative to surgery for native coarctation in infants outside the neonatal age, with low mortality, shorter hospital stay and comparable reintervention rates. In contrast, in neonates, surgery yields better results with low mortality and significantly lower reintervention rates, and should be the preferred treatment modality.

Table 1. Native aortic coarctation in neonates and infants: Comparison of outcomes with balloon coarctoplasty and surgery

Therapeutic Modality	Balloon Coarctoplasty (Group A) N = 28	Surgical Coarctation Repair (Group B) N = 47
Age (months): Median [range]	1 [0-10]	1 [0-12]
Weight (kg) (mean +/- S.D.)	3.87+/- 1.70	3.80 +/- 1.77
Sex- Male: Female	20:8	35:12
Presentation		
Shock	4	1
Tachypnea, no shock	24	46
LV dysfunction		
None	12	29
Mild	2	8
Moderate	5	5
Severe	9	5
Isthmus z-score (mean +/- S.D.)	-2.43+/- 1.47	-3.87+/- 1.63
Associated mild transverse arch hypoplasia	13 (46.4%)	22 (46.8%)
Residual coarctation at discharge	8 (28.6%)	3 (6.4%)
Hospital Stay (days): Median [range]	2 [1-20]	15 [10-44]
Follow up duration (months): Median [range]	6 [6-68]	6 [6-42]
Reintervention (neonates)	9 (60%)	4 (16%)
Reintervention (1-12 months)	3 (23.1%)	10 (45.5%)
Surgical Technique: Total (Reintervention)		
Interposition Graft		1 (0)
Pericardial patch coarctoplasty		23 (10)
Resection and end to end anastomosis		21 (4)
Subclavian flap coarctoplasty		2 (0)

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P1452 - INTERNATIONAL QUALITY IMPROVEMENT COLLABORATIVE PROGRAM IN CONGENITAL HEART SURGERY AT HOSPITAL DE NIÑOS CÓRDOBA ARGENTINA 2012 2015 EXPERIENCE

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Background: Health system experiments an increasing interest in crossing the quality care chasm between actual and expected results. Congenital Heart Surgery (CHS) has improved outcomes and quality of life in congenital heart disease; nevertheless, CHS may have infection and mortality rate that need to be sequentially quantified in order to improve results. International Quality Improvement Collaborative (IQIC) for CHS in developing countries may help to achieve these goals.
Methods: Prospective interventional study, in a tertiary public Hospital from January 2012 to December 2015. All patients with a history of CHS at Hospital de Niños de Córdoba, Argentina (HNCA) were sequentially introduced to IQIC database in order to assess: age and weight at surgery, Risk Adjust Congenital Heart Surgery (RACHS) and Infection - Mortality unadjusted rate. Key drivers were sequentially introduced as interventions: Safe Perioperative Practice, Infection Control and Team Work implementation.
Results: 373 consecutive patients were enrolled (203 males, 170 females). Annual percentage of RACHS, age and weight are shown on tables 1, 2 and 3 respectively. 2012-2015 semestral Unadjusted Infection Rate were 20%, 21.9%, 24.6%, 13.3%, 12.8%, 26%, 9.1% and 17.1% respectively being IQIC standard 4.3% -6.9% ; Semestral Unadjusted mortality rate were 6.7%, 3.1%, 6.3% ,8.9%, 11%, 0%, 0%, 4.9% respectively being IQIC standard 4,5%-6,2%. Annual Standardized Infection Rate were 1.89, 1.87, 2.0, 1.22 and Annual Standardized Mortality Rate were 0.85, 1.82, 1.07, 0.36 for years 2012, 2013, 2014 and 2015 respectively. Safe Perioperative Practice, Infection Control and Team Work implementation were sequentially introduced.
Conclusion: IQIC program implementation in CHS at HNCA contributed to quantify results and to introduce Key drivers in order to improve outcomes. Reduction in Mortality rate was successfully achieved compared to IQIC standards values and Infection rate is decreasing but still above IQIC standards.

P1455 - CONGENITAL HEART SURGERY IN LOW BIRTH WEIGHT INFANTS <2KG IN AN UNDERPRIVILIGED COUNTRY EXPERIENCE IN ARGENTINA

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Background: Low birth weight (LBW) is a risk factor for mortality in infants undergoing surgery for congenital heart disease. Typical management relies on supportive therapy or palliative surgery and delayed definitive repair. Nonetheless, the morbidity and mortality of this approach has been shown to be high. We report our experience in LBW patients (P) who underwent repair of complex cardiac defects at three institutions (Hospital Universitario Austral, Hospital Universitario Privado de Cordoba and Hospital Posadas) in Argentina.
Material and Methods: The outcomes we analyzed were hospital mortality, early reintervention, postoperative length of stay (postopLOS) and mortality (at last follow-up) in 6 P <2 kg at time of surgery who underwent open cardiac repairs from August 2008 to August 2016 at our institutions. Diagnosis were transposition of great arteries (2 P), hypoplastic left heart syndrome (2 P), aortic valve endocarditis and total anomalous pulmonary venous drainage.
Results: Average age and weight at surgery were 18.6 days (5-34) and 1.66 Kg (1.2-1.9) respectively. Surgical procedures consisted of arterial switch operation (2), Norwood (2), Ross (1) and sutureless repair of TAPVD (1). Cardiopulmonary bypass and aortic cross clamp time were 114.16 minutes (55-150) and 54 minutes (20-111) respectively. Four P had deep hypothermic circulatory arrest for 17,5 minutes (5-40). Delayed sternal closure was used in 4 P during 3.75 days (2-7). There were no unplanned early reintervention. Hospital mortality was 13%. Excluding one patient who died 20hs post surgery, postop LOS was 59.8 days (41-95). Survival was 83.3% at a mean follow up of 3.72 + /- 3.08 years.
Conclusion: Although there is risk for death in infants < 2 kg undergoing congenital heart surgery, procedures can be performed with low mortality. Our initial experience suggests that repair of complex defects is possible in LBW infants and a dedicated neonatal cardiac program yields encouraging outcomes.

P1461 - DEVELOPMENT OF AN ORTHOPEDIC APPLIANCE TO CORRECT PIGEON CHEST AFTER MIDLINE STERNOTOMY IN INFANTS

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Background/Hypothesis: ostoperative keel-type pigeon chest occasionally occurs after median sternotomy in infancy. There is no effective prophylactic method, and correction by worn appliances is required. However, current orthodontic appliances compress the entire thorax, rather than the deformity alone, and there are concerns about children wearing these appliances at important times for growth of the thorax and development of lung function. The aim of this study is to develop a new orthopedic appliance to prevent deformation and exacerbation through application of moderate compression force only to the sternal protrusion, and not to the entire thorax.
Materials and Methods: We designed a structure that has an outer shell that fits the shape of the rib cage, except for the protruding region, and a pad that generates pressure only on the protrusion. Pressure adjustment is achieved with elastic Velcro material and compression of low repulsion material. The efficacy and safety of use of this device were investigated in 18 patients, including 6 with 21-trisomy and 7 after redo sternotomy. Ages at surgery were newborn (n = 1), 1-6 months (n = 9), 7-12 months (n = 4), and 1-3 years (n = 4).
Results: The infants started wearing the appliance at 1 to 32 months (median: 11 months) after surgery and the period of use was 1 to 33 months (median: 16 months). Sagittal protrusion measured

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using a 3D scanner improved from 138 ± 8.5° before use to 151 ± 7.4° after use. Subjective evaluation of the degree of improvement by parents indicated 3 cases of improvement and 15 of prevention of progression. There were no complications such as cutaneous pressure ulcers.
Conclusions: Our orthopedic appliance improved or prevented development of pigeon chest. The most appropriate pressure setting for compression of an affected region requires data from case accumulation. Further improvements will also be made after long term follow-up.

P1462 - A NEW NON WOVEN FABRIC SHEET BASED ON SILK FIBROIN FOR USE IN CONGENITAL CARDIOVASCULAR SURGERY

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Background/Hypothesis: Expanded polytetrafluoroethylene (ePTFE) products are used in surgical correction for congenital cardiovascular anomalies, but have several drawbacks, including major bleeding from needle holes, calcification, and intra-luminal peeling resulting in stenosis. To resolve these issues, we developed a new non-woven fabric sheet with physical properties that improve surgical safety and biocompatibility. This sheet contains silk fibroin, a natural protein used in biomedicine. The aim of this study was to investigate whether the sheet is suitable for surgical use.
Materials & Methods: A solution of silk fibroin purified from silkworm cocoon and thermoplastic polyurethane (Pellethane®) was used to create a non-woven silk fibroin/polyurethane (SFP) sheet by laminated injection molding with nanofibers using the electrospinning method. The in vitro mechanical properties of the SFP sheet were compared with those of an ePTFE product. For in vivo evaluation, parts of the vascular wall of the rat abdominal aorta and the canine descending aorta were replaced with a SFP sheet or ePTFE. The sheet was explanted after 1 and 3 months for histological examination.
Results: Flexibility, water permeability, and suture retention strength of the SFP sheet were excellent and equivalent to those of ePTFE. Notably, the amount of bleeding from needle holes was negligible. The SFP patch had excellent handling properties and fitted well to the native vascular wall. The intraluminal surface of the explanted SFP sheet was covered with smooth intimal tissue without stenosis or aneurysmal changes. Histologically, there was no calcium deposition, which was detected in the excised ePTFE product, and minimal inflammatory reaction. Mild infiltration of mononuclear cells was found between the electrospun layers, but there were no signs of sheet degradation.
Conclusion: The SFP sheet had better mechanical properties and tissue compatibility than ePTFE. These favorable features and biodegradability of the SFP sheet warrant long-term follow-up.

P1472 - SYSTEMATIC REVIEW COMPARISON BETWEEN FIGURE OF EIGHT AND SIMPLE WIRE TECHNIQUE FOR STERNAL CLOSURE IN PREVENTING STERNAL DEHISCENCE AFTER OPEN HEART SURGERIES

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Introduction: Multiple techniques have been employed for sternal closure following cardiothoracic surgery. These include the figure of eight method and the simple wire technique. This study aims to investigate the advantages of employing the figure of eight technique over the simple wire method.
Objective: To assess whether the figure of eight technique is superior to the traditional technique of sternal closure in reducing the chance of sternal dehiscence in patients undergoing open heart surgeries.
Methods: In this systematic review we searched through Pubmed, Cochrane Library (Wiley) and Scopus for articles relevant to our study. We narrowed down our search to Randomized Control Trials, Clinical Trials, Retrospective Cohort Studies, Journal Analyses, Systematic Reviews and Meta-analyses published since 2000. We excluded any animal studies and articles published in any other language but English.
Results: A total of 394 articles were retrieved. Upon excluding duplications, protocols only papers, case reports, case series and reviews, 121 papers were finally selected and reviewed independently by two critics. Of the 7 articles that were finally included in the study, 4 articles stated that the figure of eight method reduced the chances of sternal dehiscence in patients undergoing open heart surgeries. 3 articles, however, showed that there was no significant difference between the incidences of sternal dehiscence with each technique of sternal closure. None of the articles showed an increase in the risk of sternal dehiscence with the figure of eight technique.
Conclusion: Majority of articles showed a decreased risk of developing sternal dehiscence with the figure of eight technique. Hence, we can conclude that the figure of eight technique is superior to the traditional wire technique since it lessens the risk of sternal dehiscence in patients undergoing open heart surgeries.

P1489 - INFERIOR CAVOPULMONARY ANASTOMOSIS IS A SAFE PALLIATION IN HIGH RISK CHILDREN ADULT FONTAN PATIENTS

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Background/Hypothesis: Superior Glenn is a standard shunt in small children with single ventricle that provides adequate pulmonary blood flow. In older children/adult with high risk Fontan, inferior cavopulmonary anastomosis (ICPA) may provide better results in term of oxygenation and bridging to Fontan.
Materials and Methods: Six patients (UVH3,CAVC2,PA-IVS1) with systemic-to-pulmonary shunt and/or pulmonary artery branches stenosis or interruption underwent ICPA using standard CPB at 33 C with extra-cardiac PTFE graft during 2005-2009. Pre and post-operative parameters were evaluated with intermediate term follow-up.
Results: Six patients (4 F,2 M) median age of 13.5 year (4.8-23.9) and weight 32 kg (13-56.6) were included. Pre-operative diagnosis were UVH + PS/PA (3cases), unbalanced CAVC (2cases) and PA-IVS (1). All except one had one or more systemic-to-pulmonary shunt. Pre-operative mean PA pressure was 20.4 mmHg (17-22), mean SpO2 was 84%(80-90). Three patients (50%) had pulmonary artery branch stenosis and one had non-confluence PA. Two patients had severe CAVC regurgitation. ICPA using PTFE graft diameter 18mm (2), 20mm (1) and 22mm (3). Standard CPB was established at 33 C (32-34) with mean bypass time of 108 minutes (90-156), mean aortic cross-clamped

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time 15 minutes (0-30). Immediate post repair mean PA pressure was 14.5 mmHg(12-17) with mean SpO2 of 90% (84-95%). There was one early death due to sudden pulmonary hypertensive crisis. Mean hospital stay was 15 days (10-24). Mean follow-up time was 3.68 year (0.55-8.06). Five patients were in normal sinus rhythm with one patient showed atrial arrhythmia. There was one late death at 14 months post ICPA. Four patients were in NYHA I with one patient was in class III. Post-operative heart size were reduced with good PA pressure in four patients (80%). Two patients had Fontan completion at 0.87 and 1.98 year after ICPA with post-Fontan follow-up of 4.28 and 5.12 year with good SpO2 (96%), both were in NYHA I with sinus rhythm.
Conclusion: ICPA is a safe palliation for children/adult with high risk Fontan. The patients can tolerate ICPA without intra-abdominal complication.

P1495 - LE COMPTE MANEUVER IN SURGICAL CORRECTION OF ABSENT PULMONARY VALVE. DOES IT IMPROVE SEVERE BRONCHIAL COMPRESSION

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Background: Here, a case of absent pulmonary valve syndrome with severe respiratory symptoms is presented. The treatment options to alleviate the airway obstruction are discussed.
Materials and Methods: A 2,5 month-old boy diagnosed of TOF and APVS was intubated because of respiratuary failure and was referred to our clinic for treatment. Pulmonary artery and the aorta were transected. Dilated pulmonary artery and branches were excised and the plicated. The maneuver of Le-Compte was then performed, translocating the pulmonary artery anterior to the aorta. End-to-end anastamosis of the aorta was completed. A size 12 valved conduit was interposed between right ventricle and main pulmonary artery.
Results: The patient was extubated shortly after readmission to the ICU. However this was short-lived because of the imminent respiratory acidosis. The chest x-ray showed total atelectasis of the left lung Thoracic CT was repeated to evaluate the bronchial tree postoperatively, which demonstrated the persisting left bronchial obstruction The details of the site and extent of the obstruction were delineated with bronchography performed by the interventional radiologist and the obstruction was dilated by deploying a 19mm (length) - 5 mm (width) balloon expandable endovascular stent.
Conclusions: Absent pulmonary valve syndrome is not only a congenital heart defect, but also a disease of the tracheobronchial system that has a high propensity to persist after the surgical correction of the heart defect. Regardless of the surgical technique used, the persisting respiratory disorder may complicate the postoperative course and may result in adverse outcomes. Although its main disadvantage is the lack of progressive grow in children, endobronchial stent appication may be an effective option in such cases.

P1505 - ESTABLISHMENT OF A COMPREHENSIVE BLOOD CONSERVATION PROGRAM REDUCES BLOOD UTILIZATION ESPECIALLY FOR JEHOVAH'S WITNESS PATIENTS

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Background: Cardiac surgery on Jehovah's Witnesses (JW) can be challenging given the desire to avoid blood products. Establishment of a blood conservation program involving the pre-, intra-, and post-operative stages for all patients may lead to minimized need for blood transfusion in all patients.
Methods: Pre-operatively all JW patients were treated with high dose erythropoietin 500 U/kg twice a week. JW patients were compared to like non-JW patients from the congenital cardiac database, two per JW to serve as control. Blood use, ventilation time, bypass time, hematocrit pre-operative, first in intensive care unit (ICU), and at discharge, and 24-hour chest drainage were compared. Preoperative huddle, operating room huddle and postoperative bedside handoff were done with the congenital cardiac surgeon, perfusionist, anesthesiologist, intensive care team in all patients for goal alignment.
Results: Five JW patients (mean weight 24.4 24.9 Kg, range 6.3–60 Kg) were compared to 10 non-JW patients (weight 22.9 22.2 Kg, range 6.9–67.8 Kg). There was no difference in bypass, cross clamp, time to extubation (0.8 versus 1.7 hours), peak inotrope score (1.8 versus 2.0), or chest drainage. No JW patient received blood product compared to 30% of the non-JW. Pre-operative Hct was statistically greater for the JW patients (46.1 3.3% versus 36.7 4.6%, $p < 0.001$), and ICU and discharge Hct were also higher for the JW (37 1.8% vs 33.8 7.4%, and 41 8.1% versus 32.8 7.4%). All patients had similar blood draws during the hospitalization (JW x 5 mL/admission vs. non-JW 6 mL/admission).
Conclusion: Developing a comprehensive blood conservation program for congenital heart patients requires teamwork that includes goal alignment, by using erythropoietin and supplemental vitamins we could perform all JW surgeries without any blood products. Our overall program has minimized blood product administration.

P1508 - HYPERCARBIC CEREBRAL BLOOD FLOW RESERVE IN SINGLE VENTRICLES PRIOR TO AND AFTER BIDIRECTIONAL GLENN PROCEDURE

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Background/Hypothesis: The cerebral and systemic vascular beds vasodilate while the pulmonary vasculature vasoconstricts in response to hypercarbia; this is a sign of cerebrovascular health and intactness. We hypothesized that in large group of single ventricle (SV) patients, the increase in cerebral blood flow (CBF) under hypercarbic conditions of patients prior to bidirectional Glenn (pre-BDG) will be less than those of BDG (cerebral and pulmonary vascular beds in series) because the higher systemic runoff of pre-BDG physiology will limit the increase.
Materials/Methods: SV patients were studied with cardiac magnetic resonance phase contrast mapping in the jugular veins immediately pre-BDG and after BDG in both room air and in hypercarbic conditions (3-7 torr of carbon dioxide administered over 20 minutes).
Results: 170 consecutive subjects were studied: 58 pre-BDG (4.8 + /-1.7 months) and 112 BDG (2.9 + /-1.4 years). From room air to hypercarbia, significant increases in CBF were demonstrated at both the pre-BDG and BDG stages (both with $P < 0.05$) of 35-40% – see graph. CBF in BDG were significantly higher than pre-BDG in both room air and hypercarbia ($P < 0.05$), however, the amount of CBF increase during hypercarbia

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from room air within each group was not statistically significant (P = 0.18). Using a mixed effects linear regression model, CBF increased by 0.596 liters/minute/meter2 after adjusting for stage.
Conclusions: Robust increases in CBF in SV during hypercarbia both prior to and after BDG occurs. The incremental CBF increase in response to hypercarbia within each group is not different, indicating the systemic runoff of pre-BDG does not limit the increase. Hypercarbic CBF reserve is therefore similar in both groups, however, it should be noted that CBF of BDG is higher than pre-BDG in both room air and during hypercarbia. As CBF has been implicated in neurodevelopment, this finding may have implications for neurodevelopmental outcomes.

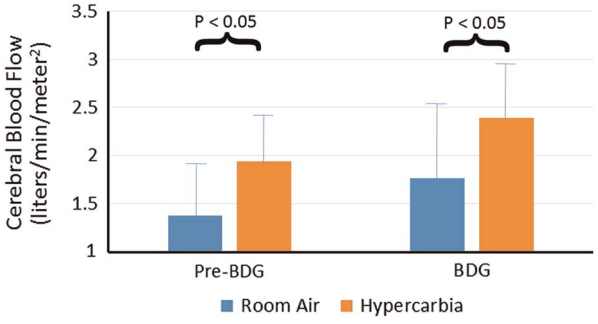

Figure.

P1511 - REDUCING THE INCIDENCE OF ACUTE KIDNEY INJURY (AKI) MULTIDISCIPLINARY TEAM APPROACH AND GOAL ALIGNMENT IN A CONGENITAL HEART PROGRAM

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Background: Acute Kidney Injury (AKI) has been shown to be associated with increased morbidity and mortality in pediatric congenital cardiac surgery. AKI has been shown to occur in adult and pediatric post-operative cardiac surgical patients between 18 and 60%. The purpose of this study was to evaluate the incidence of AKI comparing the pediatric risk, injury, failure, loss, end (pRIFLE) and the acute kidney injury network (AKIN) scoring systems in a new congenital heart program.
Methods: Perfusion and anesthesia guidelines were established to minimize fluid administration and hemodilution, and included pre-operative goal alignment. Retrospective analysis of all congenital heart surgeries requiring cardiopulmonary bypass between 2014 and 2016. Values for serum creatinine measured pre-operative (baseline), first on arrival to the intensive care unit (ICU), twenty-four, and forty-eight hours, urine output for the first 24 hours, bypass time, ventilation time, re-intubation, and length of stay were pulled from the quality database.
Results: Eighty cardiopulmonary bypass cases were performed, but 4 cases were excluded for pre-operative renal failure leaving seventy-six bypass cases. 28% (n = 11) had an AKI by the pRIFLE and 14.5% (n = 11) by the AKIN. The primary difference between the two scores was the requirement of a 0.3 mg/dL increase in serum creatinine with the AKIN plus a 1.5x increase over baseline. None of the patients met the oliguria guideline and none of the patients required renal replacement therapy. By the AKIN score 2.6% were stage 1, 9.2% stage 2, and 2.6% stage 3. Average Inotrope score on arrival to the ICU, peak 48 hours, length of stay, and ventilator time all increased with each increase in score.
Conculsion: Low AKI scores are achievable in a new congenital cardiac surgery program and when AKI occurs is associated with an increasing inotrope score may help guide clinical care.

P1515 - UNBIASED PROFILING OF THE GENE EXPRESSION CHANGES THAT OCCUR IN NEONATES EARLY DURING CARDIOPULMONARY BYPASS

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Rationale: The systemic inflammatory response seen during the first 24 hours after patients undergo cardiopulmonary bypass (CPB) represents a significant clinical problem, especially in neonatal patients. Unfortunately, the molecular pathogenesis of this process is poorly understood. Understanding the gene changes that occur during CPB would advance efforts to limit the post-CPB inflammatory response, thereby improving outcomes for these patients.
Methods: RNA was isolated from the nucleated blood cells before CPB and 1 hour after the initiation of CPB from neonatal patients who underwent cardiac surgery. The five patients (3 males, 2 females) were between 6 and 19 days of age at the time of surgery. 3 of the patients had total anomalous pulmonary venous return. One had a diagnosis of transposition of the great arteries. The other patient had double inlet left ventricle. mRNA sequencing was performed on these samples. Bioinformatic analyses were performed with a multiple testing corrected p-value of $p < 0.05$ being considered significant.
Results: 1230 genes were altered 1 h after the initiation of CPB (496 genes were upregulated). There was significant enrichment of genes associated with cytokine response ($p = 8.32 \times 10\text{-}12$) with 8.1% of the genes changed being associated with this gene ontology group. 7.6% of the genes changed were associated with gene ontology group for innate immunity ($p = 2.0 \times 10\text{-}9$). There was significant enrichment for the transcription factors NFAT ($p = 1.06 \times 10\text{-}9$), EGR (early growth response factor) ($5.46 \times 0\text{-}8$), and NF-kB ($p = 6.84 \times 10\text{-}4$) within 5 kilobases of the start sites for the genes at were changed 1 hour after the initiation of CPB.
Conclusions: To our knowledge, this abstract is the first report of unbiased profiling of the gene changes that occur during first hour of CPB in neonatal patients. Our bioinformatic analyses suggests that modulation of NFAT, EGR, and/or NF-kB signaling may result in amelioration of the post-CPB inflammatory response.

P1520 - LONG TERM SURVIVAL AFTER EXTRACORPOREAL LIFE SUPPORT FOLLOWING PEDIATRIC CARDIAC SURGERY A STUDY FROM THE PEDIATRIC CARDIAC CARE CONSORTIUM

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Background: Extracorporeal Life Support (ECLS) is increasingly used during recovery after surgery for congenital heart diseases (CHD).

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Although successful to salvage critically ill patients in the immediate postoperative course, the long-term outcomes of ECLS survivors after operations for CHD are not well studied. We used a multicenter US-based registry, the Pediatric Cardiac Care Consortium, to examine the long-term survival and risk of transplant of patients with history of ECLS after pediatric cardiac surgery.
Methods: We identified 348 children needing ECLS after surgery for CHD between 1982 and 2003. Patients that survived to discharge were linked with the National Death Index (NDI) and the United Network for Organ Sharing (UNOS) using direct identifiers to determine their long term vital and transplant status until 12/31/2014.
Results: A total of 117 patients (34%) were discharged alive, 87 with two-ventricle and 30 with single-ventricle physiology. Follow up data through NDI and UNOS were provided for a median of 13.8 years (max 24.9 years) after discharge. The 15-year transplant-free survival was significantly higher in patients with two-ventricle physiology compared to those with single-ventricle (76% vs. 43%, $p < 0.001$)(see Kaplan-Meier plot). The median time of death or transplant after discharge from the ECLS-associated operation is 15.8 years for the two-ventricle and 8.4 years for the one-ventricle group respectively. Single-ventricle patients included 22 with stage I physiology (9 Norwood and 13 aorto-pulmonary shunts); of them 14 (63.6%) underwent Glenn procedure with 10 (45.5%) reaching Fontan completion, while 8 (36.3%) needed transplant or died without additional CHD surgery.
Conclusions: ECLS survivors after CHD surgery for two-ventricle lesions have an acceptable long term transplant-free survival. For single-ventricle patients, history of ECLS support is associated with short survival after discharge even though 2/3 of them will be able to progress to at least one of the subsequent.

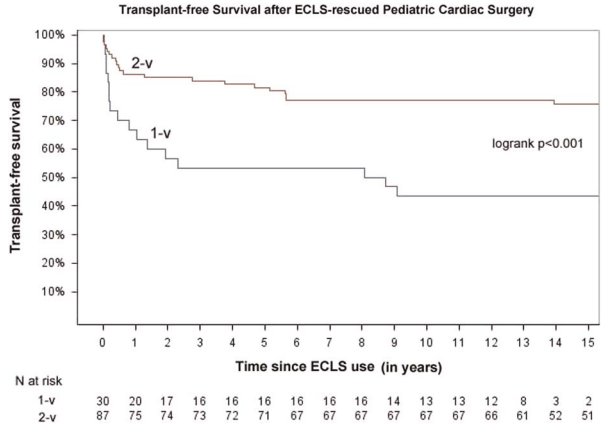

Figure.

P1521 - EARLY AND RAPID STENOSIS OF AORTIC POSITION MITROFLOW®LXA BOVINE PERICARDIAL TISSUE VALVE – A FOLLOW UP STUDY

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Background/Hypothesis: We previously reported that young patients who underwent aortic valve replacement (AVR) with the Mitroflow® LXA, compared with Magna®/Magna Ease®, bovine pericardial valve were at high risk for early failure and rapid progression to severe prosthetic aortic stenosis. We here present the longer-term follow-up of an expanded group of patients.
Materials and Methods: All patients with Mitroflow® LXA and Magna®/Magna Ease® AVR were followed for prosthetic valve function by serial echocardiograms.
Results: AVR with the Mitroflow® LXA was performed in 18 patients (44% male) at median age 18.9 years (range 7.6-40.0), and with the Magna®/Magna Ease® in 63 patients (56% male) at median age 20.0 years (range 3.8-66.7). Freedom from failure with the Mitroflow® LXA valve vs. Magna®/Magna Ease® valve, respectively at 1, 2, 3, and 5 years was 100% vs. 100%, 77% (95% confidence interval [CI] 50-91%) vs. 100%, 48% (CI 23-68%) vs 100%, and 26% (CI 8-50%) vs. 94% (CI 63-99%) (logrank $p < 0.001$). Failed Mitroflow® LXA valves experienced a period of hyper-accelerated stenosis with progression from ≤ mild to severe stenosis over a 3-12 month period despite absence of symptoms. In contrast, Magna®/Magna Ease® valves were explanted for severe stenosis and endocarditis in one patient each, 6.1 and 3.1 years post implant, respectively. Five Mitroflow® LXA valves remain in situ ≥ 3 years in patients 20-40 years old at implant; 2 have severe structural valve deterioration.
Conclusions: With longer-term follow-up, the Mitroflow® LXA valve continues to demonstrate premature failure in young patients, particularly those under 30 years of age at implant. Rapid progression from mild to life-threatening aortic stenosis occurred over months. Young patients should not undergo AVR with the Mitroflow® LXA valve, and those with the valve in situ require heightened echocardiographic surveillance due to rapid development of stenosis.

P1524 - SURGICAL OUTCOME OF BILATERAL PULMONARY ARTERY BANDING FOR BIVENTRICULAR REPAIR

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Objective: Recently bilateral pulmonary artery banding (BPAB) is performed for avoiding preoperative risks of the congenital heart defects depending on patent ductus arteriosus even if it was originally performed for TAC. We select BPAB if patient had a risk such as shock status, low body weight (<2.5 kg) or severe extra cardiac anomalies. The purpose of this study was to investigate the outcome of BPAB for biventricular repair (BVR).
Patients and Methods: 10 patients underwent BPAB between 2003 and 2015. There were IAA/CoA in 7 and TAC in 3. BPAB was performed by lasso technique.
Results: Median age and body weight at the surgery were 7.2days (1-27 days), 2.6 kg (1.6-3.2 kg), respectively. The reasons of BPAB were shock in 6, low body weight in 5, LVOT size in 2, ventricular size in 1 and extra cardiac anomaly in 1. The circumference of BPAB was 12.5mm (9.0-13.5 mm). The duration to 2nd surgery was 62 days (28-313 days). Re-PAB (adjustment) was required in 2. Completion of BVR was in 7 (Yasui in 3, Rastelli in 2, ICR in 2), Waiting of BVR in 2, HD in 1 (preoperative brain hemorrhage due to shock). One patient died at 5 years after Rastelli operation. 2 patients with low body weight (1.6 kg, 1.8 kg) required PTA before 2nd operation because of decreased pulmonary blood flow. 6 patients required PA angioplasty or PTA at BVR or later.

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Summary: The reason of BPAB was simply avoiding risk rather than morphological reason. BVR for low body weight patients could be performed after several PTAs. Although PA angioplasty was required at BVR or conduit replacement, there was no re-operation for just branch PS because PTA was effective.

P1527 - LONG TERM OUTCOMES AFTER SURGERY FOR ANOMALOUS LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY A STUDY FROM THE PEDIATRIC CARDIAC CARE CONSORTIUM

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Background: Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiovascular anomaly with limited information regarding its long-term outcomes after surgical intervention.
Methods: This is a cohort study from the Pediatric Cardiac Care Consortium, a multi-institutional US-based registry of interventions for congenital heart diseases. We identified 186 patients with the diagnosis of ALCAPA having adequate identifiers for linkage studies. Long term outcome data are provided by linkage to the National Death Index (NDI) and the United Network for Organ Sharing (UNOS) up to the latest update of 12/31/2014.
Results: The median age at time of repair was 6 months (range: 9 days-17.2 years). Of the 186 patients, 141 underwent coronary re-implantation (76%), 28 Takeuchi procedure (15%), 7 coronary artery bypass grafting (4%), and 10 ligation of the anomalous coronary artery (5%). A total of 162 patients survived to discharge (87%). Concomitant procedures at time of repair for these individuals included mitral valve surgery (n = 14), interatrial communication closure (n = 11), and patent ductus arteriosus ligation (n = 3). Twenty-four patients (14.8%) needed subsequent procedures including mitral valve surgery (n = 9), coronary artery intervention (n = 7), pulmonary angioplasty (n = 10), heart transplant (n = 4) and other procedures (n = 5). There were 8 deaths (two after heart transplant) over 32 years of follow up as provided by linkage to the NDI and UNOS. The 20-year transplant-free survival rate was 94.3% for ALCAPA patients discharged alive (see Kaplan-Meier survival plot), with a median follow-up of 17.9 years (IQR = 14.3-23.4).
Conclusions: 20-year transplant-free survival after surgery for ALCAPA reaches 94.3%, but there is significant remaining morbidity requiring additional interventions. The most common long-term complications after surgical intervention are mitral regurgitation, Takeuchi tunnel leak or stenosis, pulmonary artery stenosis and heart failure.

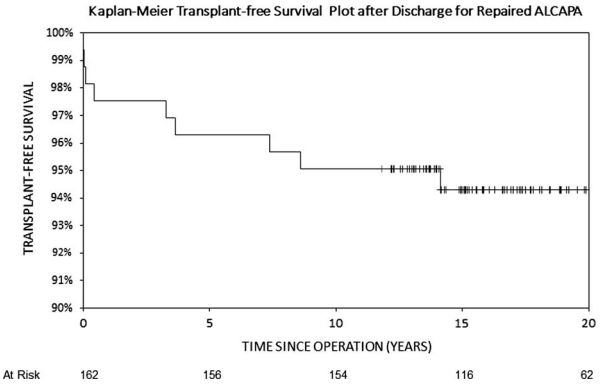

Figure.

P1528 - CLINICAL SIGNIFICANCE OF LEFT VENTRICULAR HYPERTROPHY IN INFANTS AFTER VENTRICULAR SEPTAL DEFECT REPAIR

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Background: Left ventricular hypertrophy (LVH) is a not uncommon post-operative change after ventricular septal defect (VSD) repair in infants. The mechanism and clinical effect of this post-operative LVH are still unclear.
Methods: From 2013 to 2015, all consecutive patients received surgical repair of isolated VSD within 12 months of age were enrolled. We analyzed the pre- and post-operative echocardiographic data (within 14 days after surgery) and correlate with clinical parameters.
Results: Totally 37 infants (24 male and 13 female) were enrolled. The mean age of operation was 3.49 months, with mean duration of ICU stay and hospitalization 6.16 ± 4.69 and 15.4 ± 7.74 days respectively. Increase in LV mass was observed in 13 (35.1%) patients (LV thickening group). Their mean LV mass Z-score was +0.40 (-3.38 to +2.99) before surgery and +1.69 (-1.38 to +4.88) after surgery. We compare demographic data, pre-operative parameters, bypass time, cooling temperature, and post-operative clinical course between two groups. LV thickening group had smaller LVEDD Z-score (+1.18 vs +4.60, $p = 0.005$) and LV mass index (70.2 vs 127.5 g/m2, $p < 0.001$) pre-operatively. Cooling temperature during operation tends to be lower in LV thickening group (28.5°C vs 30.5°C, $p = 0.062$), but not bypass time and aortic cross clamp time. As for post-operative clinical course, there's no significant difference in duration of intubation, inotropic agent use, ICU and hospital stay between two groups, but chest tube drainage duration tends to be longer in LV thickening group (6.23 vs 4.50 days, $p = 0.058$). The LVH regressed at later follow-up and normalized within 3 months.
Conclusions: LVH after VSD repair occurs in a significant portion of infants with isolated VSD. It implicates with smaller pre-operative LV size, LV mass index, and lower operative cooling temperature. However, the clinical course is benign and will not prolong hospitalization.

P1535 - INFERIOR CAVOPULMONARY ANASTOMOSIS WITH SIMPLE TRICUSPID ANNULOPLASTY IS EFFECTIVE TREATMENT FOR SEVERE FORM EBSTEIN'S ANOMALY

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Background/Hypothesis: Repairing of severe forn Ebstein is technically demanding and provided unpredictable results in unexperience hands. Young surgeon may need long learning curve due to its rare incident. Inferior cavopulmonary anastomosis (ICPA) with simple tricuspid valvuloplasty (STV) might be a simple and reproducible technique to manage these rare problems.
Materials and Methods: Fourteen patients with severe Ebstein underwent ICPA with STV using standard CPB at 32 C during 2005-2014. ICPA performed by using extracardiac PTFE graft.

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Tricuspid valve repair by simple annuloplasty, suturing clefts or closure of TV. Associated lesions included ASD (11),PS/PA + PDA(2),WPW(3) were also corrected. Pre and post-operative clinical parameters were evaluated. Statistical data represented in mean with range.
Results: 14 patients (F = 9, M = 5) with severe Ebstein (type C = 12, type B = 2), mean age 16y (5.4-48y), mean weight 38.4 kg (14.2-83 kg) were included. All were symptomatic (cyanosis = 11, dyspnea = 7, palpitation = 3, CHF = 2, fainting = 1). Standard CPB was established at 32 C. ICPA using extracardiac PTFE graft diameter 16mm (1), 18mm (5), 20mm (5) and 22mm (3). Tricuspid valvuloplasty by simple annuloplasty (5), suture cleft (8), closure (1). Intra-operative alasion of WPW was done in 1 patient. Mean bypass time was 103 minutes (87-155), mean aortic cross-clamped time was 33 minutes (0-64). All patients were in NYHA I post-operatively vs pre-op NYHA II (4), III (7), IV (3). Post-op CT ratio reduced from 0.57 (0.44-0.78) to 0.52 (0.42-0.68). Post repair echo showed no TR (6), mild TR (6), moderate TR (1) vs pre-op mild TR (2), moderate TR (4), severe TR (8). Post-op SpO2 improved to 96.9% (89-100%) vs pre-op 88% (73-96). Post ICPA + STV CVP was 9.2 mmHg, mean PAP was 14.8 mmHg (12-22) vs pre-op 14 mmHg (13-15). CCU stay was 3.5 days (1-9). ICD drainage was 2.4days (1-4). Hospital stay was 10.6 days (4-20). There was one early death on day2 due to low cardiac output. Follow-up the patients (n = 13) to 27.99 patient year (0.2-11.3y). No late death. No reoperation. The first case still alive 11.3y post surgery in NYHA I without reoperation/medication.
Conclusion: ICPA + STV is safe and reproducible technique to treat severe Ebstein. Improvement of functional class, degree of TR, oxygenation and reduce cardiac size were demonstrated.

P1545 - ASSESSMENT OF CHILDREN WITH COMPLETE VASCULAR RING

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Backgraound: A complete vascular ring is a rare congenital cardiovascular anomaly which encircles the trachea and oesophagus compressing them. This study assessed the presentation symptoms, diagnostic methods and treatment results after surgery in children with complete vascular ring.
Methods: Symptomatic complete vascular ring patients undergoing surgery between January 2010 and August 2016 at Ege University, Faculty of Medicine Hospital,İzmir, were retrospectively assessed. The presentation symptoms, demographic characteristics, and diagnostic tests were evaluated. Operative data and postoperative follow up, complications and problems were also examined in detail.
Result: Nine patients underwent surgery, 6 (66%) of whom were male. Median age was 10 months (range, 28 day- 4 years). Among these patients, 44,4% (n = 4) had ligamentum arteriosum, 55,5% (n = 5) had double aortic arch. Three patient had right-sided aortic arch + ligamentum arteriosum, One patient had left-sided aortic arch + aberrant right subclavian artery, and Kommerell's diverticulum was identified in three patient. Preoperative tracheomalacia was identified via bronchoscopy in two patient. Concomitant tracheal reconstruction was not performed in any patient. Computed tomography (CT) angiography was performed in 5/9 patients. All the patients were successfully operated on. None patient died in the early period.
Conclusions: Outcomes of division of a complete vascular ring are excellent. Preoperative bronchoscopy, oesafography and CT angiography is effective for the differential diagnosis and visualization of vascular ring abnormality. Complete vasculer ring couse of ligamentum arteriosum treat with transected both of the ligamentum arteriosum and fibrous cord. In such cases, full recovery can be assured with early diagnosis and surgery.

P1549 - MINIMALLY INVASIVE TECHNIQUE OF PLACING A DUAL CHAMBER PERMANENT PACEMAKER IN CHILDREN

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Background: Permanent pacing in pediatric population is challenging with consideration of body growth, patient's size, presence of coexisting congenital heart disease and intra-cardiac shunts. There are different approaches for permanent pacemaker implantation which includes thoracotomy, sternotomy or Video Assisted Thoracotomy. We present a technique for dual chamber permanent pacemaker implantation via the xiphisternal approach.
Materials and Methods: This is a retrospective review of all cases of dual chamber pacemaker implantation using the xiphisternal approach performed at the Aga Kahn University from January 2011 to December 2016.
Operative Procedure: The patient is placed in a supine position. A 4-6 cms vertical midline incision over the xiphoid process is made to gain entry into the pericardial cavity. A pericardial well is created. The atrium is held with soft clamps. Atrial pacing leads are attached to the body of the right atrium using two 5/0 polypropylene. Ventricular pacing leads are implanted on the diaphragmatic surface of the right ventricle using two interrupted 5/0 polypropylene sutures each. The pacemaker generator box pocket is created beneath the rectus abdominis muscle via a transverse incision in the left lumber region. The pacing leads are connected to the generator via a subcutaneous tunnel. After maintenance of a satisfactory pacing threshold by an electro-physiologist the anterior rectus sheath, subcutaneous tissue and skin are closed. The pericardium is left open and the xiphoid incision is closed in layers.
Results: This technique was successful in 15 patients, (3 months - 10 years), in both primary and post-cardiotomy patients with no morbidity. Successful atrio-ventricular synchrony was established along with excellent pacing and sensing thresholds. This technique is associated with minimal surgical trauma, pain and results in early discharge.
Conclusion: This minimally invasive Xiphisternal technique of implanting a dual chamber pacemaker in children is easily reproducible and associated with minimal morbidity.

P1565 - THE ROLE OF SURGICAL VALVULOPLASTY IN PEDIATRIC PATIENTS WITH CONGENITAL AORTIC STENOSIS

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Objectives: Balloon valvuloplasty is firmly established as effective treatment of congenital aortic stenosis (AS) in children. Surgical techniques of aortic valve repair have improved and there is controversy on the best approach to treat congenital AS. Direct surgical valvotomy with exact splitting of fused commissures, shaving off of obstructing nodules and enlarging of aortic sinuses may produce a better functioning valve in comparison to blind ballooning.

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Methods: Retrospective review of data from 31 consecutive patients (boys = 26, girls = 5, newborns = 8) undergoing surgical valvuloplasty for isolated congenital valvular AS as the initial interventional procedure between 2012-2015. Median (IQR) age at surgery was 8.6 months (34 days–5.1 years). Median (IQR) clinical and echocardiograhic follow-up was 1.6 (0.8-2.9) years. Aortic regurgitation was graded as 0 = none, 1 = mild, 2 = moderate and 3 = severe.
Results: Prior to surgery the median diameter (z-score) of the aortic annulus was 11.2 mm (-1.24 z). Operation included aortic valvotomy in all patients and additional procedures in 29/31: cusp shaving = 15, enlargement of aortic sinus using autologous pericardium = 15, pericardial cusp extension = 5 and left ventricular outflow tract myectomy = 2. No patient has died. One patient had to be re-operated. Peak aortic systolic gradient was efficiently reduced by surgery. Aortic regurgitation was well tolerated and remained to be less than moderate in the majority (Table).
Parameter Before surgery After surgery Last follow-up p.
Peak systolic aortic gradient [mmHg].
median (IQR) 100 (90-108) 34 (28-45) 44 (31-57) <0.001.
Aortic regurgitation [grade].
median (IQR) 0 (0–0.5) 1.0 (0–1.5) 1.0 (0.5–2.0) <0.001.
Conclusions: Surgical valvuloplasty remains our strategy to treat patients with congenital AS if we can offer more than simple valvotomy. The procedure is safe. Short- to mid-term reoperation free survival and functional results are favorable (Supported by MH CZ - DRO, University Hospital Motol, Prague, Czech Republic 00064203).

P1567 - USE OF DUAL AORTIC CANNULATION TECHNIQUE FOR AORTIC ARCH REPLACEMENT IN CHILDREN WITH AGGRESSIVE GENETIC AORTOPATHIES

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Background: Children with genetic aortopathies are at risk of developing aneurysmal dilation of the aortic arch following ascending aortic replacement. The value of aortic arch replacement at the time of initial surgery has been debated with opponents citing increased complexity and risk of adverse outcome due to prolonged deep hypothermic circulatory arrest (DHCA). At our institution, total aortic arch replacement is performed with the use of a dual cannulation technique (innominate artery and descending aorta) with continuous mildly hypothermic full-flow cardiopulmonary bypass (CPB) to the entire body. We sought to compare surgical outcomes in children with genetic aortopathies undergoing either the traditional valve-sparing aortic root replacement (VSRR) or VSRR with total arch replacement with use of DAC (AAR-DAC).
Methods: Record review of all children (age <21) with a genetically confirmed diagnosis of aortopathy undergoing aortic surgery in the past 5 years was performed. Patients undergoing concomitant valve surgery or Ravitch procedure were excluded.
Results: VSRR was performed in 7 patients and VSRR plus total arch with DAC in 3. There was no difference in age (14.4 + /-5.8 vs 8.5 + /5.8, p = 0.17), weight (57.7 + /-28.9 vs 50.7 + /-49.6, p = 0.79), CPB time (166.3 + /-49.0 vs 205.0 + /-59.9; p = 0.31) or length of stay (4.9 + /-1.9 vs 4.3 + /-1.5, p = 0.68). Six of the 7 patients undergoing VSRR and 2 of 3 undergoing AAR-DAC were extubated in the OR.
Conclusions: Despite a more complex operation, patients undergoing additional aortic arch replacement with use of DAC and avoidance of DHCA had no increase in complication rate with mean time to discharge of only 4 days. Consideration of aortic arch replacement at the time of initial VSRR should be considered in patients with Loeys-Dietz syndrome and other aggressive aortopathies.

P1572 - ARCH REPAIR IN UNUSUAL AORTIC CONFIGURATION OTHER THAN LEFT AORTIC ARCH AND LEFT DESCENDING AORTA

Jong Yoon Park[1], Jeong-Jun Park[1], Won Kyoun Park[1], Bo Sang Kwon[2], Chun Soo Park[1], Tae-jin Yun[1], Jae Suk Baek[2], Jeong Jin Yu[2], Young-hwue Kim[2], Jae-Kon Ko[2]
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Background: One of major concerns during arch repair is to prevent airway compression. Majority of arch obstruction patients have a configuration of left arch and left descending aorta. However, some patients have unusual configuration which may cause more challenging surgical technique and a postoperative airway compression. We reviewed our experiences of arch repair in these patients retrospectively since December 2010.
Methods: Among the total 198 arch repaired patients, nine patients with unusual aortic configuration other than left arch and left descending aorta underwent arch repair after CT evaluation. Body weight was 3.1 ± 0.5 kgs. Aortic arch obstruction was coarctation of aorta in 8 (including 2 HLHS variant) and type B interrupted aortic arch in 1. Aortic configuration were circumflex retroesophageal left aortic arch in 2, left aortic arch and right descending aorta crossing vertebrae in 5, right aortic arch and left descending aorta in 1, and right aortic arch and right descending aorta with retroaortic innominate vein in 1. In eight patients, aortic arch was repaired under selective cerebral perfusion after extensive mobilization of descending aorta. One patient who had no intracardiac defect underwent arch repair via left thoracotomy.Five patients needed patch material for arch reconstruction including two Norwood operations. Intraoperative bronchoscopy was done routinely.
Results: There was no mortality. There were no additional operations for arch stenosis or airway compression. All patients achieved the aortic configuration at the same side of arch and descending aorta.Two patients on the single ventricle repair tract finished Fontan operation following neonatal Norwood operation.
Conclusion: Individualized arch repair even in unusual aortic configuration provides a good midterm result and preoperative CT scan is very helpful for the surgical planning. Also, extensive mobilization of descending aorta is mandatory and using patch material is helpful.

P1577 - COR TRIATRIATUM WITH TRANSPOSITION OF GREAT ARTERIES VSD UNROOFED LSVC IN A YOUNG ADULT – A RARE COMBINATION RESULTING IN PHYSIOLOGIC ATRIAL SWITCH CASE REPORT AND SURGICAL SIGNIFICANCE

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Cor triatriatum represents 0.1% of all congenital cardiac malformations and may be associated with other cardiac defects in as

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many as 50% of cases. The association of Cor with transposition of great arteries has not been reported before. We report, how a rare combination of anomalies lead to physiologic atrial switch leading to his late presentation and how with altered simple surgical approach, significant palliation could be obtained, in this rare combination of anomalies. 22 yr old presented to us with gradually worsening dyspnea on exertion over the last 5-6 yrs, when he presented to medical attention 5 yrs back he was informed that he has inoperable complex congenital cardiac problem. He came to us with worsening dyspnea over the last 1 month, he was saturating 80% in room air with significant clubbing. He had a hemoglobin of 20 g%. The RA incision was like superior septal approach for the mitral valve. The right atrial appendage was sutured to the apex of the incision at the roof of left atrium. This resulted in a wide opening so that the pulmonary veins could drain unimpeded into the right atrium At one year follow up he is saturating 90% and is class NYHA class I. This case is reported for (1)Rare association of Cor triatriatum with TGA and VSD (2) How the Cor membrane functions the first layer of Senning's procedure and how in this patient, the presence of unroofed LSVC and occluded RSVC by aneurysmal PA and atrial septal defect resulted in physiologic Sennings procedure.

P1584 - CLINICAL AND FINANCIAL IMPACT OF ON TABLE EXTUBATION AFTER OPEN HEART SURGERY IN CHILDREN – A RETROSPECTIVE COHORT STUDY

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Background: On-table extubation after open heart surgery in children is beneficial in a developing country. It is cost effective, reduces hospital stay, increases patient satisfaction with a low complication rate.
Objective: To determine the impact of on-table extubation on morbidity, mortality and cost benefit in pediatric patients undergoing open heart surgery.
Study Design: This is a descriptive retrospective cohort study. The study population includes all open heart surgeries performed at Aga Khan University in children (6 months -18 years) from January 2012 – June 2014.
Methods: All patients with Risk Adjustment for Congenital Heart Surgery category 1-3 were included while those in 4-6 were excluded. Criteria for extubation; patients fully concious, hemodynamically stable and with normal post-operative arterial blood gases and echocardiogram. Complications included re-intubation, cardiopulmonary arrest, high chest tube output, respiratory acidosis and low cardiac output syndrome. The patients were anesthetized using a standard technique and the patients were assessed for OTE using standard extubation criteria.
Results: Out of 630 surgeries, 108 patients (15.4%) underwent OTE. Surgical correction included Ventricular Septal Defect, Tetralogy of Fallot, Atrial Septal Defect, Glenn shunts and Fontan surgery. There were 7 complications. One patient needed re-intubation due to respiratory failure, one patient had atrial arrhythmia, another patient had respiratory acidosis, two required tube thoracotomy for pleural effusions and two had superficial sternal wound infections. There was no mortality. The estimated cost of an intubated and ventilated patient is PKRs: 60,000/= while a non-intubated patients cost is 30,000/=. The length of stay of an extubated patient in CICU is less than 24 hours and hospital stay lesser by 48 hours as compared to an intubated patient.
Conclusion: OTE is an attractive practice with positive impact on cost and has low complication rates in a selected group of pediatric patients undergoing open heart surgery.

P1600 - TECHNICAL PERFORMANCE SCORE AS A PREDICTOR OF PERIOPERATIVE OUTCOMES IN COMPLEX CONGENITAL HEART SURGERY PERFORMED IN A LOW VOLUME SURGICAL PROGRAM

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Objective: Technical Performance score (TPS) has been associated with postoperative outcomes in large volume surgical programs. We assessed the ability of TPS to predict perioperative outcomes in complex congenital heart surgeries performed in a low volume program.
Methods: Eighty-eight consecutive patients undergoing cardiac surgery over 4 years at our institution were retrospectively reviewed. A TPS was assigned to each patient based on discharge echocardiographic findings and need for re-interventions in anatomic areas of interest.
Results: There were 76 STAT 4 and 12 STAT 5 category patients. Fifty-five patients were neonates (62%), 19 (22%) infants, 10 (11%) children and 4 (5%) adults. Sixty percent of the patients were males and 16% were premature. An optimal TPS score (class 1) was assigned to 56 (64%) patients. An adequate TPS score (class 2) was assigned to 13 (15%) patients and an inadequate TPS score (class of 3) was assigned to 11 (13%) patients. Eight patients were not scored as their operation was not included in the original TPS scoring system. Median length of stay (LOS) was 16 (IQR: 9, 29) days. 30-day and in-hospital mortality rate was 6% ($n = 5$). Early post-op complications (within 30-days post discharge) included: delayed sternal closure (26%), reintubation (17%), arrhythmias (17%), chylothorax (10%), pneumothorax (4%), infection (3%), cardiac re-operation (1%), and stroke (1%). No patient required dialysis post-discharge. Unplanned readmission rate was 14% ($n = 12$). TPS score was associated with reintubation, ICU LOS and post-op LOS ($p < 0.05$). Other outcomes such as unplanned 30-day readmissions, reoperation, inotropic score and mortality were not shown to be effected by TPS score ($p > 0.05$).
Conclusions: Technical performance score is a useful tool in predicting postoperative outcomes after high complexity cardiac surgery in a low volume program and should be part of program review and quality improvement initiatives in congenital heart programs of all sizes.

P1602 - INCIDENCE ETIOLOGY AND RISK FACTORS OF UNPLANNED READMISSIONS AFTER CONGENITAL HEART SURGERIES

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Objective: The first year after congenital heart surgery (CHS) is a period of increased health care utilization. We analyzed the rate, etiology, risk factors and patient mortality associated with unplanned readmissions within one year following CHS.

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Methods: We retrospectively reviewed 262 patients undergoing CHS at our institution from 08/2011-06/2015. Scheduled readmissions including second stage palliation for single-ventricle patients were excluded.
Results: Seventy patients (27-infants, 21-children, 22-neonates) were readmitted 119 times (1.7times/patient). On multivariate analysis, age, STAT-category, surgery and intubation time, ICU and hospital length-of-stay (HLOS) were risk factors associated with unplanned readmissions. Leading causes of unplanned readmissions within 12-months post-CHS included viral illness (34%), respiratory distress (16%), gastrointestinal (14%) and cardiac causes (8%; Table-1). Forty-six (39%) of these readmissions were within 30-days post-discharge. Leading causes of 30-day readmissions included viral illness (33%), wound infections (15%) and cardiac causes (13%; Table 1). Median time-to-first readmission was 21 (IQR: 12-58) days. Median HLOS at readmission was 2 (IQR:1-8) days. The 1-, 3-, 6- and 12-month freedom from first readmission was 83%, 73%, 71% and 69% respectively. There was no significant difference in survival between readmitted and non-readmitted patients ($p = 0.68$). Time-to-readmission and readmission frequency didn't impact patient survival ($p > 0.05$).
Conclusions: Unplanned readmissions are common during the first year after CHS especially in high risk patients. Viral infections were the leading cause of unplanned readmissions. Readmissions didn't impact long-term survival of patients after CHS.

Table 1. Major primory etiologies of readmission after congenital heart surgery.

Readmission Etiology	No. of Readmissions	No. of Patients	No. of Readmission/ patient	No. of 30-day readmissions
Viral illness	40	26	1.5	15
Respiratory distress	19	16	1.2	4
Gastrointenstinal	17	14	1.2	4
Cardiac	10	10	1.0	6
Neurological	9	8	1.1	1
Superficial wound infections	8	8	1.0	7
Other	8	7	1.1	4
Infection other	5	5	1.0	2
Pericardial/ pleural effusions	3	3	1.0	3
Total	**119**	**70**		**46**

Other = chylothorax, failure to thrive, anemia, weight loss.
Infection other = otitis media, urinary tract infections, infectious cellulities.

P1626 - RESULTS OF VALVE SPARING AORTIC ROOT REPLACEMENT SURGERY IN ASIAN POPULATION A SINGLE CENTER STUDY

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Objective: We aimed to investigate the outcomes of valve sparing aortic root replacement surgery (David procedure) in patients with connective tissue disease in Asian population.
Method(s): All patients underwent David procedure were retrospectively reviewed. Patient's presentation, severity of aortic root dilatation and post operative outcomes were examined.
Results(s): Between January 2013 and December 2016, 8 David procedure was performed in National Heart Centre Singapore. 7 patients have been diagnosed of Marfan syndrome and 1 with Loeys-Dietz syndrome (4 Male, 4 Female, age 11-51). 5 patients had elective operation performed for dilated aortic root during follow up; 3 patients presented with Type A dissection and had emergency David procedure performed, 2 of emergency cases also had concurrent ascending aorta and aortic arch replacement with elephant trunk. Mean pre-operative sinus of valsalva size was 5.14 cm (3.9 cm – 6 cm, 3.9 cm with Z score of +9.8 belonged to the 11 years old Loeys-Dietz syndrome patient). There were 2 in-hospital mortalities, both patients had cardiovascular collapse post emergency David procedure and concurrent surgeries. All other patients had smooth recovery and still on follow up. All patients, including the 2 demised patients, had post operative echocardiogram showed no more than mild degree of aortic regurgitation with acceptable gradient. Mean follow up was 512 days; cumulative follow up was 8.4 patient-year.
Conclusion: Valve sparing aortic root surgery is an effective treatment for aortic root dilatation with connective tissue disease, sparing patients from disadvantages of prosthetic valve. In our experience, patients with just David procedure do much better than those who had more complex surgeries (David procedure + surgery on other part of aorta) done. Longer duration of follow up is needed to assess aortic valve competency in long term.

P1634 - NORWOOD OPERATION WITH PRIMARY CAVOPULMONARY SHUNT IN UNIVENTRICULAR HEARTS WITH HIGH FLOW UNRESTRICTED PULMONARY BLOOD FLOW

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Objective: Children with univentricular heart lesion with coarctation and arch hypoplasia are treated with a Norwood operation with a systemic to pulmonary shunt as the first stage of palliation. Delayed presentation to specialised centers is a challenge in developing countries. A subgroup of patients who present late and have high pulmonary blood flow and low resistance may be treated by a primary cavopulmonary shunt (CPS) as a source of pulmonary blood flow at the time of the Norwood operation.
Methods: Patients with univentricular heart lesions including hypoplastic heart syndrome (HLHS) who presented beyond the neonatal period without previous interventions and with evidence of low pulmonary resistance were treated with a Norwood type operation combined with a primary bidirectional CPS. Patients received postoperative pulmonary vasodilators and were followed through hospital discharge and as outpatients.
Results: Seven patients received the Nrowood-CPS operation. Four patients had HLHS, one with mitral atresia and aortic stenosis, and two with mitral stenosis and artic stenosis. One of the latter patients also had an interrupted inferior vena cava. Two patients had tricuspid atresia with transposed great arteries and coarctation of the aorta. One patient had double inlet left ventricle with coarctation and arch hypoplasia. All patients had unrestricted pulmonary blood flow with systemic pressure in pulmonary arteries. Six patients underwent preoperative heart catheterisation showing reactive pulmonary vasculature with left to right shunt. The age at operation ranged from 6 weeks to 1 year. Six patients survived the operation and were discharge, operative mortality was 14%. One patient, who was 1 year of age at the Norwood-CPS operation, died of pulmonary hypertension and failed cavo-pulmonary circulation.

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Conclusion: In late presenters requiring a Norwood type operation, primary CPS is a viable option if there is evidence of low pulmonary vascular resistance.

P1639 - IN HOSPITAL INTERSTAGE CAN DECREASE MORTALITY AFTER NORWOOD STAGE ONE OPERATION THE BROMPTON'S EXPERIENCE

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Objective: interstage mortality after Norwood stage-one remains between 12 and 20% in current series. We present our preliminary experience with in-hospital interstage program, aimed at facilitating escalation of care, possibly improving survival until stage-two.
Methods: a prospective study was designed for patients with diagnosis of HLHS and HLHS-variants, offering in-hospital stay after Norwood operation until stage-two completion. Daily and weekly investigational plans were tailored to the individual patient. Primary end-point was overall survival until stage-two.
Results: between May 2015 and August 2016, 15 consecutive patients with prenatal diagnosis of HLHS (9) or HLHS-variants (6), underwent Norwood stage-one (13) or comprehensive-Norwood operation (2). Preliminary bilateral pulmonary artery banding was performed in 7. Stage-one Norwood-Sano was preferred in 11 (73.3%). In-hospital interstage was achieved in 14 patients (93.3%). Norwood operative and interstage survival were 93.3% and 100% at 20 months (mean follow-up 9.8 ± 2.2 months). During in-hospital interstage, 5 patients (33.3%) required urgent and efficacious escalation of care, due to haemodynamically compromising atrial arrhythmias affecting tricuspid valve function (2), or Sano shunt stenosis (3). These potentially fatal events occurred 2.4 months after primary Norwood-Sano. Additional interstage interventions included: residual aortic arch obstruction repair (1) neo-aortic valve repair (1), redo Sano (1) and Sano-conduit dilatation (1) or stenting (1) by catheter intervention. Bidirectional Glenn (10) or comprehensive-Norwood stage 1&2 (2) were successfully completed in 12 patients at 5.1 months with 100% survival. Two patients are currently awaiting stage-two within the in-hospital interstage plan.
Conclusions: in-hospital interstage program facilitates escalation of care, which is possibly efficacious in reducing interstage mortality after Norwood stage-one.

P1640 - RISK FACTORS FOR PERSISTENT PLEURAL EFFUSIONS FOLLOWING TOTAL CAVOPULMONARY CONNECTIONS

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Background: Total cavopulmonary connection (TCPC) is followed by significant fluid retention manifesting commonly as pleural effusions. We aimed to study various pre, intra and postoperative factors including serum cortisol in producing persistent pleural effusion defined as lasting for more than two weeks or more than 20 mL/kg/day.
Materials and Methods: We conducted a prospective cohort study of 38 patients undergoing TCPC between September 2015 and November 2016. All patients underwent trans thoracic echocardiography and cardiac catheterization. Factors studied included age, weight, height, symptomatology, echocardiography findings of normal or diseased atrio- ventricular valves, aortic and pulmonary valves, ventricular function, pulmonary artery pressures, oxygen saturation in the femoral artery, presence of aorto pulmonary collaterals, type of TCPC procedure, cardiopulmonary bypass duration, inotropic requirement, mechanical ventilation, conduit size and fenestration size, if any.
Results: The mean age at operation was 13.1 ± 5.6 years. The mean duration of chest tube drainage was 15.76 ± 13.2 days and the mean drainage was 9.15 ± 4.6 mL/kg/day. The statistically significant risk factors for pleural effusions were history of syncope ($p = 0.04$), pulmonary artery (PA) pressures ($r = 0.328$, $p = 0.003$, odds ratio 1.30), higher hemoglobin levels ($p = 0.04$), Wernowsky inotropic score ($r = 0.4$, $p = 0.01$), rise in the serum cortisol ($p = 0.03$) and central venous pressure at 6 hours ($r = 0.44$, $p = 0.005$) and 12 hours ($r = 0.4$, $p = 0.01$) in the post operative period. The duration of mechanical ventilation showed a statistically significant positive correlation with the amount of pleural drainage ($r = 0.45$, $p = 0.005$). Although the study showed a positive correlation of Fontan pressure with pleural drainage but a statistical significance could not be obtained ($r = 0.25$, $p = 0.13$). There was 1 in- hospital mortality following Fontan failure and takedown.
Conclusions: Higher PA pressures (>15 mmHg), higher Wernowsky inotropic score, higher hemoglobin levels, higher CVP and lower serum cortisol levels post operatively were associated with persistent pleural effusions after the TCPC.

P1651 - RIGHT VENTRICULAR OUTFLOW TRACT RECONSTRUCTION USING A HAND SEWN TRILEAFLET VALVE CONDUIT

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Background: Prohibitive cost and subsequent unavailability of valved homograft or CONTEGRA conduits lead us to construct our own hand-sewn conduits for right ventricular outflow tract (RVOT) reconstruction. We have implanted hand-sewn conduits made of a bovine pericardial tube with 1mm PTFE (PRECLUDE/ GORE TEX) trileaflet cusps. This report assesses the short-term outcome of these prostheses.
Material and Methods: From 2012 to 2016, self-sewn tri-leaflet conduits were implanted in 46 patients. During surgery on a back-table a piece of bovine pericardium is taken and cut to size as per a pre-calculated nomogram. A 1mm PTFE membrane is similarly cut and shaped to pre -determined size and is then sewn onto the bovine pericardium as three semilunar leaflet cusps. This is then sewn into the shape of a tube, thus making a valved conduit.
Results: The mean age at operation was 9.6 years (range: 0.1 to 38 years). The most common procedure performed was a Rastelli operation in 28 patients followed by Ross procedure in 10 patients. 4 patients had previously placed conduits replaced and 2 patients had primary Truncus arteriosus repair, while 2 had primary repair of Tetralogy of Fallot (TOF). The most common size conduits were 20mm (24 patients) and 22mm (19 patients). Follow-up was complete. There were 2 in-hospital deaths. Both were unrelated to the conduit. None of the 46 PTFE tri-leaflet

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conduits developed significant obstruction. Mean gradient across the valve conduit was 19 mmHg 10-38 mmHg). No patient has required re-operation for graft failure till last follow up There was one late death in a patient who developed infective endocarditis in the implanted conduit. Overall functional status of patients was excellent at last follow-up.
Conclusions: Short term results show that hand-sewn tri-leaflet conduits have provided a reliable conduit for RVOT reconstruction. Longer follow-up is needed to determine durability and longevity.

P1665 - ENDOPHENOTYPIC PATHWAYS DETERMINE THE ANATOMIC VARIANTS OF ATRIOVENTRICULAR SEPTAL DEFECTS

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Background: An atrioventricular septal defect (AVSD) is characterized by a common AV junction. Its anatomic variants are defined by the associated defects of AV septal components. Genetic mutations are known that cause AVSD in general, but the pathways that lead to an anatomic variant are undefined.
Methods: We quantified the incidence of AVSD caused by Nkx2-5 haploinsufficiency in four crosses involving five inbred mouse strains (1293 to 8406 hearts/cross; 14,723 total). Anatomic endophenotypes of the atrial and ventricular septae were examined. The incidences of AVSD, anatomic variants and endophenotypes were compared between crosses to detect the effect of genetic modifiers. Correlations between endophenotypes were assessed to infer the hierarchy of developmental pathways that lead to the anatomic variants of AVSD.
Results: AVSD incidence in Nkx2-5 + /- hearts varied from 0.4-7.6% between the crosses. No AVSD was found in the wildtype. All anatomic variants were observed, even unbalanced AV canal defects and ones with an intact atrial and ventricular septum. The incidences of certain variants varied significantly between crosses. An absent vestibular spine is a hallmark of an AVSD, yet one was present in 20-40%, depending upon the cross. Hypoplastic vestibular spines were common and appeared related in part to insufficient growth of the dorsal mesenchymal protrusion in either the anterior-posterior or apical-basal axes. Deficiencies of the inlet ventricular septum could be classified as either anterior or posterior. The former is correlated with a "divot" on the right ventricular side of the left ventricular outflow tract septum. The latter is correlated with an absent vestibular spine.
Conclusions: Our results support a model in which an AVSD mutation causes a common AV junction, while polymorphic modifier genes in endophenotypic pathways determine the susceptibility to AVSD in general and the anatomic variant. Correlated defects of certain AV septal components suggest novel pathway relationships.

P1666 - MID TERM OUTCOMES OF TRANSVENTRICULAR VALVOTOMY APPROACH FOR INITIAL TREATMENT OF PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM

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Objective: To review the medium term surgical outcomes of transventricular pulmonary valvotomy(TPV) in patients with PA/IVS and non-right ventricular dependant coronary circulation.
Methods: Data were reviewed retrospectively for 21 patients with PA/IVS treated with TPV from 2010-2013 in our center. The mean body-weight of the 21 patients was(3.2 ± 0.7)kg and mean age was(16 ± 8)d. The Z-score of tricuspid valve ranged from -2.5~2.2(-0.9 ± 0.4). All the patients were followed up for over 12months. All the patients were divided into two groups: Bi-ventricle repair(BVR) and non-BVR group and the percutaneous saturation, Z-score of tricuspid valve, reintervetion data were collected at 3,6,12,24,36,48 months after the procedure.
Results: There was one early death after the TPV procedure for severe hyoxemia and parents refused to reoperation. 12 patients finally underwent BVR and 5 underwent one and half ventricle repair. 3 patients were on follow-up after TPV. The mean follow-up time was (32.3 ± 10.3)months without late death. Early reintervention were needed in 4 patient(20%). One was in BVR group and accepted the percutaneous pulmonary valve balloon dilation for residual obstruction. 3 were in one-BVR group and got hyoxemia need modified B-T shunts in 2 and PDA stent in 1 patient (8.3% vs 37.5%, $p < 0.01$). Z-score in BVR group was significantly larger than non-BVR group, with mean Z-score of (-0.5 ± 0.3) and (-1.5 ± 0.6), respectively($p = 0.01$). The freedom from reoperation was 80, 70, 35% at 6 months, 1 and 3 years, respectively. In BVR group, 9 of 12 received the ASD closure. In non-BVR group, 5 received bidirectional Glenn shunt and close the ASD; 1 received reconstruction of right ventricle outflow for muscular obstruction. The mean Z-score ranged from -2~2.2 (-0.5 ± 0.4),-2.1~2.2(-0.5 ± 0.3), -2.3~2.0(-0.2 ± 0.3), -0.4~2.2 (0.5 ± 0.8)~-0.5~2.2(0.6 ± 0.8), -0.2~2.2(0.7 ± 0.3) at 3months, 6months,1year,2,3 and 4years after TPV.
Conclusions: The results of a hybrid approach to treating patients with PA/IVS are excellent and proved to be safe and effective to improve the function of right ventricle. TPV can be used as an alternative method to transcatheter approach.

P1669 - ATRIOVENTRICULAR VALVE RECONSTRUCTIONS USING LEAFLET EXPANSION AND A SUB PARTIAL ANNULOPLASTY

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Background: Atrioventricular valve reconstruction in the pediatric population is a challenge due to the frequent combination of type I and IIIb dysfunction, i.e. annular dilatation and leaflet restriction. The need for growth potential further challenges the available repair techniques. We have elected a strategy using leaflet expansion and sub-partial annuloplasty with PTFE reinforcement.
Materials and Methods: From August 2014 through October 2016, six children aged five months – three years (median 19 months) underwent elective atrioventricular valve repair due to severe regurgitation (MR/TR) at Lund University Hospital, in Sweden. Their primary diagnoses were idiopathic congenital mitral regurgitation (three patients), progressive mitral regurgitation after previous AV canal repair (one patient), severe mitral regurgitation after treatment for endocarditis (one patient), and hypoplastic left heart syndrome with depressed right ventricular function and severe secondary tricuspid regurgitation. Their surgery consisted of leaflet expansion with autologous, untreated pericardium and sub-partial annuloplasty with PTFE reinforcement. Two patients had one additional procedure performed each: VSD closure and ductus ligation.
Results: All children survived the surgeries with uneventful post-operative courses, except for one patient who needed an early

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reoperation for functional stenosis due to a "spinnaker phenomenon", which was corrected by creating neo-chordae with PTFE. The sub-partial annuloplasty was not performed in this patient, being the first in this serie. All patients were alive and asymptomatic after a median follow-up of eight months (range one – 22 months). Their echocardiographic data at that time showed no greater than 1 MR/TR and a median gradient of 4 mmHg (range 3-9 mm Hg).
Conclusions: Leaflet expansion with autologous pericardium and sub-partial annuloplasty with PTFE reinforcement for atrioventricular regurgitation in the pediatric population gives satisfactory results both early and at intermediate follow-up. However, the long-term results and growth potential needs to be studied further.

P1672 - CAROTID ARTERY RECONSTRUCTION WITH CRYOPRESERVED CAROTID HOMOGRAFT

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Background: Carotid artery reconstruction is rare in the pediatric population. Moreover, the need for growth potential further limits the surgical options. We describe two cases were we have used cryopreserved homografts for carotid artery reconstructions.
Materials and Methods: The two patients were operated on in November, 2015 and July, 2016, respectively. The first patient was 10 years old and had progressive bilateral carotid artery and aortic arch stenosis as components of a supravalular aortic stenosis syndrome. He underwent carotid artery bifurcation plasty with an aortic homograft patch at the origin of the right common carotid and subclavian arteries with support of a innominate artery -carotid artery shunt and subsequent patch enlargement of his entire aortic arch including the first two centimeters of the left carotid artery using a branched aortic arch homograft. The latter surgery was performed with extracorporeal circulation, deep hypothermia, and circulatory arrest. The second patient was a 10 year old boy with a tracheostomy due to nemalin myopathy. He had a severe bleeding from a tracheo-innominate artery fistula that was initially treated with endovascular stenting. When the bleeding recurred, the patient underwent excision of the innominate artery and the proximal two centimeters of the right common carotid artery and the subclavian artery, respectively, excising all the infected stent material and replacement by an adult-sized homograft consisting of innominate-carotid-subclavian arteries. The operation was performed with extracorporeal circulation and moderate hypothermia.
Results: Both patients had an uneventful postoperative course with no signs of perioperative cerebral injury. Echocardiographic evaluation revealed unobstructed laminar flow. Both patients were asymptomatic at follow-up after 5 and 14 months, respectively.
Conclusions: Cryopreserved homografts are a valuable vascular graft for carotid reconstruction. However, we have only used it in two patients and the the long-term results and growth potential needs to be studied further.

P1673 - POSTOPERATIVE FOLLOW UP OF PATIENTS WITH ATRIOVENTRICULAR SEPTAL DEFECT AT BASKENT UNIVERSITY BETWEEN 1996 2016

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Purpose: Follow-up results of patients with atrioventricular septal defect (AVSD) operated in 1996-2016 at Başkent University are presented.
Method: Data obtained from hospital records consists of echocardiographic and angiographic details before surgery, age and weight at surgery, operative details, presence of Down's syndrome, details of postoperative care (durations of mechanical ventilation, inotropic agent infusion, stay in intensive care unit-ICU, inotropic score, postoperative arrhythmias, infections, time to discaharge), early postoperative and latest echocardiographic findings and hospitalization for reintervention.
Results: 498 patient-files were reviewed including 335patients (67.4%) with complete and162patients (32.6%) with partial AVSD (48.6% Down syndrome). AV valve morphology was Rastelli A in 90% (299), B in 9% (29), C in 1% (4) of patients. All patients underwent biventricular repair. Operative technique was single-patch in 21.7% (108), double-patch in 26% (129) and modified single-patch technique (Wilcox) in 52.3% (260). Left AV valve cleft was unsutured in 27 patients. Follow up time was 37.82 ± 46.77 (range 0.5-96) months. 68patients (12%) had arrhythmias in ICU, pacemaker was implanted in 6patients. 78patients (17%) were treated for pulmonary hypertensive crisis. Early-mortality-rate was 9.7 (47patients). In late follow-up, 11patients underwent left AV valvuloplasty, 16patients had MVR. Left ventricular outflow tract obstruction (LVOTO) was seen in 19patients. Late mortality was due to postoperative death after MVR in 2patients. Late reintervention was more frequent in modified single-patch technique ($p = 0.04$). Patients with Down syndrome were younger and weighed less at surgery; duration of mechanical ventilation, frequency of infection and hypertensive crisis were more; late LVOTO and AV-valve reintervention were less. Severe left AV valve regurgitation was more in patients without Down syndrome.
Conclusion: Morbidity and mortality rates are still high early after surgery in AVSD. Although Down syndrome patients are disadvantaged in early postoperative period, late morbidity and mortality are less.

P1674 - A NOVEL TECHNIQUE FOR PATIENTS WITH SCIMITAR SYNDROME WITHOUT ASD AND A SMALL LEFT ATRIUM – THE "TRENCH TECHNIQUE"

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Background: The partial anomalous pulmonary venous connection seen in Scimitar syndrome is a challenge to reconstruct, especially in the setting of an intact atrial septum and a small left atrium. The surgical correction carries a relatively high risk for the development of a subsequent tunnel obstruction. We describe a novel technique that we have used in two patients with Scimitar syndrome.
Materials and Methods: From July, 2015 through September, 2016 two children, aged five months and three months, underwent elective surgical correction of their partial anomalous pulmonary venous connection. Both children had a complete venous drainage from their right lung via a single vertical scimitar vein that descended to enter the inferior vena cava posteriorly. Neither of them had an atrial septal defect and both left atria were small. One

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of the patients was highly symptomatic with heart failure, severe pulmonary hypertension and failure to thrive. The other patient had only mild pulmonary hypertension and was asymptomatic. The technique consisted, apart from the traditional intracaval tunnel formation and atrial septectomy, of creating a posterior widening of the tunnel by a full thickness posterior incision and expansion by autologous pericardium (one patient) and cryopreserved pulmonary homograft patch (one patient). This "trench"widened the tunnel and expanded the left atrium, thereby reducing the angle of venous return from the right lung to the left atrium.
Results: Both patients had an uneventful postoperative course. There were no signs of obstructions of the pulmonary venous return at follow-up(two and 16 months, respectively) and the children were asymptomatic.
Conclusion: The "trench technique" for anomalous pulmonary venous connection seen in Scimitar syndrome seems to be a promising strategy. However, we have only used it in two patients and the long-term results and growth potential needs to be studied further.

P1685 - REGIONAL DIFFERENCES IN COST AND LENGTH OF STAY AMONG NEONATES WITH HYPOPLASTIC LEFT HEART SYNDROME

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Background: Hypoplastic left heart syndrome (HLHS) is one of the most resource-intensive congenital heart diseases. Understanding geographic variation in mortality, hospital charges and length of stay (LOS) can inform policy decisions and aid quality improvement efforts.
Methods: Infants ≤28 days old and born with HLHS between 2000-2012 were identified using the Kids' Inpatient Database. Hospitalizations were stratified into two groups: 1) birth hospitalization and 2) secondary/transfer hospitalizations. United States regional differences in hospital charges and LOS were compared using adjusted linear regression.
Results: 2,431 primary birth hospitalizations were identified. There were 449 (18.5%) inpatient deaths, with significant differences across regions ($p = 0.02$). Infants in the Northeast had the shortest LOS and lowest hospital charges. Compared to the Northeast, LOS in the South was, on average, 7.8 days longer (95% CI 2.8, 12.9,$p = 0.002$), but no significant differences in LOS in the Midwest and West were seen. The South also charged, on average, $93,483 more (95% CI $29,145, $157,820), compared to the Northeast, the Midwest charged $77,983 more (95% CI $4,682, $151,284) and the West $171,275 more (95% CI $99,492, $243,059). 1,895 secondary hospitalizations were identified, with 472 (24.9%) inpatient deaths. Significant differences in mortality were observed across regions ($p = 0.02$). Compared to the Northeast, the South stayed, on average, 10.4 days longer (95% CI 4.9, 15.9,), the Midwest 7.0 days longer (95% CI 1.6, 12.4), and no significant differences were seen in the West. Additionally, the South charged, on average, $212,398 more (95% CI $141,228, $283,567), the Midwest $139,617 more (95% CI $70,098, $209,136), and the West $103,302 more (95% CI $35,565, $171,039).
Conclusions: Significant geographic variations in mortality, LOS, and hospital charges exist in care of HLHS newborns in the US. A better understanding of these differences could improve quality of care and reduce costs..

Table 1. Standardized average length of stay (LOS) and hospital charges (in thousands of dollars), stratified by region, among neonates during primary hospitalizations, n = 2,431, and secondary hospitalizations, n = 1,895.

	Average LOS (95% CI)	Average Hospital Charges[a] (95% CI)
Primary hospitalizations[b]		
Northeast	24.9 (20.8, 29.0)	272.4 (220.4, 324.4)
Midwest	28.5 (24.4, 32.5)	350.4 (298.7, 402.0)
South	32.7 (29.9, 35.5)	365.9 (330.0, 401.8)
West	28.5 (24.8, 32.2)	443.7 (395.1, 492.3)
Secondary hospitalizations[b]		
Northeast	29.0 (24.9, 33.1)	274.0 (220.5, 327.5)
Midwest	36.0 (32.8, 39.2)	413.6 (372.1, 455.0)
South	39.4 (35.8, 43.1)	486.4 (438.8, 533.9)
West	27.2 (24.1, 30.3)	377.3 (337.7, 416.9)

[a]In thousands of dollars
[b]Adjusted for admit year, gender, race/ethnicity, insurance status, household income, comorbidities, surgical procedures, placement of a gastronomy tube, inpatient mortality, hospital size, and hospital teaching status

P1698 - OUTCOMES FOLLOWING MECHANICAL ATRIOVENTRICULAR VALVE REPLACEMENT IN PATIENTS UNDERGOING SINGLE VENTRICLE PALLIATION

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Background: Outcomes following mechanical atrioventricular valve (AVV) replacements in patients with single ventricles are uncertain.
Methods: From 1997-2016, 14 consecutive patients with functionally single ventricles underwent mechanical AV valve replacement for valve dysfunction at our institution. Operative mortality was defined as death within 30 days of surgery or prior to hospital discharge.
Results: Seven patients (50%) had hypoplastic left heart syndrome, 4 (29%) had an unbalanced atrioventricular septal defect, 2 (14%) had tricuspid atresia and 1 patient (7%) had pulmonary atresia with intact ventricular septum. Prior to AVV replacement, 12 patients (85%) had qualitatively normal or mildly reduced ventricular function and 2 (14%) had severely decreased function. 13/14 (91%) had severe AVV regurgitation. Median age of replacement was 2 years old (range 0.36- 6.75 years). St. Jude's valve was used in all patients (range 19 to 31mm). Following AVV replacement, 6 (43%) patients had normal or mildly reduced ventricular function, 3 (21%) had moderately decreased function and 2 (14%) patients had severely reduced ventricular function. Operative mortality was 43% (6/14). There were 2 late deaths. Kaplan-Meier survival estimates of the cohort are shown in Figure 1. Follow up was 100% complete with a median follow up period of 5.4 years (range, 0.08-8.8 years). 4 (21%) patients had complete heart block postoperatively that did not recover. 6 (43%) patients reached Fontan completion, 2 of whom died. Significant bleeding occurred in 57% (8/14) with 3 patients suffering major cerebrovascular accidents. There were 5 events of valve thrombosis in 4 patients. At last follow up, 2 patients had moderate or severe ventricular dysfunction, 1 had severe valve stenosis and none had significant valve regurgitation. No independent risk factors for mortality were identified.

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Conclusion: Mechanical valve replacement carries significant mortality and morbidity. Careful consideration should be made before embarking replacement.

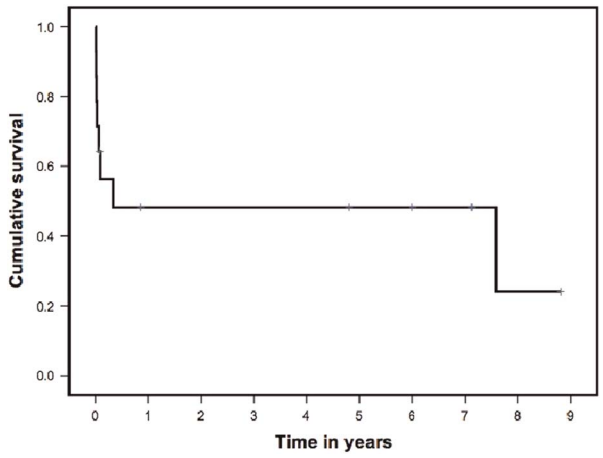

Figure.

P1702 - MID TERM FOLLOW UP OF TRICUSPID VALVE ANATOMICAL RECONSTRUCTION (CONE REPAIR) IN EBSTEIN'S ANOMALY

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Ebstein's anomaly (EA) is a congenital heart defect (CHD) in which the septal leaflet of the tricuspid valve (TV) is displaced into the cavity of the right ventricle toward its apex. Its prevalence is 0.5-1% of all congenital heart defects. In some cases the defect is diagnosed late because primarily patients are asymptomatic, but the final stage of this disease is a heart failure. Usually, a first symptoms are serious arrhythmias or sudden death.
Objective: To analyze a mid-term follow up of successful cone reconstruction of TV in Ebstein's anomaly.
Methods: From 2012 to 2016 in Ukrainian Children's Cardiac Center 26 patients with Ebstein's anomaly were operated by performing the cone reconstruction of TV. The mean age of patients was 7.8 + 3.2 years. Preoperative diagnosis was made by the anatomy of the tricuspid valve: type "A" in 5 patients, type "B" in 10 patients, type "C" in 8 patients, type "D" in 3 patients. All patients were conducted electrophysiological study (if necessary, conducted high-frequency catheter ablation) and a cardiac catheterization before surgery. 7 patients had cone reconstruction combined with the Glenn shunt because of right ventricle failure.
Results: Early postoperative mortality was 7.7% (2 patients). Mid-term follow-up was 26.5 + 14.8 months. During the follow-up visit the clinical condition of the patients was examined, tests of physical tolerance, ECG, echocardiography, chest X-Ray and magnetic - resonance imaging of the heart were performed. By Echo there were from mild to moderate insufficiency on the TV, contractile function of the left ventricle is good and mild decreased right ventricle's contractility, indicating good result of correction. In late postoperative period reoperation was performed in 3 patients through a severe tricuspid insufficiency.
Conclusions: Cone reconstruction of TV is a very effective technique of surgical anatomical treatment of Ebstein's anomaly with good follow up results.

P1711 - AIR EMBOLISM TEAM TRAINING; NEEDS ASSESSMENT USING SIMULTION FOR PEDIATRIC CARDIAC SURGERY

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Introduction: Massive air embolism (MAE) is a rare but potentially catastrophic event. A recent root-cause analysis of a MAE event identified communication as a contributing factor. Recognizing patient safety is a collective function of individual clinicians, multidisciplinary teams and organizational systems we report on utilization of high fidelity simulation team training for MAE response.
Method: 44 clinicians were divided into 7 teams, each containing a pediatric cardiac surgeon, anesthesiologist, nurse/scrub technician and perfusionist. The MAE scenario was performed twice; bi-caval and single atrial cannulation techniques, as well as pre and post aortic cross clamp periods to ensured typical evaluation. Team performance was assessed by expert raters for gaps in procedural skills and team communication. Each scenario was debriefed with intention to address identified performance gaps. Learners' perceived value of simulation-based training was also solicited.
Results: Observations indicated poor team communication was primary barrier to successful execution of MAE protocol. Post-evaluation participant responses were positive. All participants felt the training program should be made mandatory, and 95% of respondents suggested at least annual training. Sample responses included, "This training was great. It gives you the opportunity to know what to expect should this occur," suggesting nurses and scrub technicians felt they had a better understanding of the protocol.
Conclusion: Comprehensive team training using high fidelity simulation for MAE has potential to improve understanding of procedure and protocol. Evaluation of the program's impact on cardiac team communication should be assessed prior to full implementation.

P1725 - PULMONARY VALVE REPLACEMENT IN PATIENTS WITH RIGHT VENTRICLE OUTFLOW RECONSTRUCTION

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Background: The most frequent reoperation is the pulmonary valve replacement in patients having performed right ventricle outflow reconstruction. In this article, the early surgical outcome of the patients who had undergone PVR was discussed.
Materials and Methods: Seventy-six patients had PVR surgery for pulmonary insufficiency that developed after right ventricle outflow reconstruction between 2009 and 2016. Symptoms like

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exercise intolerance or arrhythmias that are attributed to right ventricular volume overload or dysfunction with or without branch pulmonary artery stenosis were considered as surgical indications for PVR. Surgical indication was made according to MRI measurement in asymptomatic patients.
Results: There was no early mortality and late mortality after 60 ± 19 months of follow up. The mean right ventricular ejection fraction (RVEF) value in symptomatic patients is increased in the postoperative term, although this result is statistically insignificant. There is decrease in right ventricular area index (RVAI), right ventricular volume index (RVHI) and tricuspid annulus measurements compared to preoperative results.
Conclusions: PVR can be performed with low morbidity and mortality rates. It shall be performed before the irreversible changes in the right ventricular volumes and functions begin.

P1727 - SHOULD EXTUBATION IN OPERATING ROOM BE THE STANDARD OF CARE IN TETRALOGY OF FALLOT REAPIR EVIDENCE FROM A PROSPECTIVE OBSERVATIONAL STUDY

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Background: Despite known benefits early extubation is not widely practiced in tetralogy of Fallot (TOF) repair. Aim of study was to assess feasibility of extubation in operating room in TOF repair and identify factors hindering early extubation strategy.
Methods: This was a prospective observational study between January 2015 and September 2016 which included 132 consecutive patients undergoing total correction for TOF. Extubation in the operating room was aimed for in all cases. Data was collected for baseline characteristics and operative variables to assess outcome measures. Risk factors were analyzed to identify independent correlation with failure to extubate in operating room.
Results: 132 (69 males, 63 females) patients underwent total correction for TOF. 77(58.3%) were extubated in the operating room. Of the remaining 55(41.7%) the median ventilation time was 105 minutes (inter quartile range: 71-135 mins.). 25/132 (18.9%) patients were less than 10 kg in body weight and 13/132 (9.8%) patients were less than one year in age. Transannular patch was created in 87/132(65.9%) while pulmonary valve was preserved in 45/132(34.1%) patients. Mean cardiopulmonary bypass time was 86.5 ± 20.3 mins and mean aortic cross clamp time was 57.5 ± 14.7 mins. Mean ICU stay was 23.1 ± 4.4 hrs and average post operative length of stay was 4.8 ± 0.6days. There was one death (1/132, 0.7%) due to low cardiac output. None of the risk factors analyzed appeared to be related to a failure to extubate on-table- age <1 year(Odds ratio 0.81, 95% CI 0.25-2.57); weight <10 kgs (OR 0.72; 95% CI 0.3-1.74); post repair Right Ventricle/ Left Ventricle (pRV/LV) ratio > 0.7 (OR 1.01; 95% CI 0.43-2.34); cross clamp time (OR 0.72; 95% CI 0.35-1.42); CPB time (OR 0.68; 95% CI 0.31-1.5).
Conclusions: Most children undergoing total correction for repair of tetralogy of Fallot can be extubated safely in the operating room. None of the perceived risk factors are real impediments for extubation in operating room.

P1730 - SURGICAL MANAGEMENT FOR PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM: A SINGLE CENTER 17 YEARS EXPERIENCE AND MEDIUM TERM FOLLOW UP

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Objective: The optimal surgical strategies for pulmonary atresia with intact ventricular septum (PAIVS) are still not well established. This study reviewed our 17 years' experience for the management of PAIVS.
Methods: Between July 1999 and June 2016, 194 patients were treated for PAIVS in our heart center. Based on the morphology of right ventricle (RV), age and surgical approaches, the patients were divided in to two groups: one-stage surgery group (n = 39) and staged surgery group (n = 155), in which received definitive repairs, including biventricular repair, 1.5 ventricular repair and univentricular palliation without or with initial intervention. Median follow-up time was 6.9 years (range, 1 to 17 years), and survival, risk factors for death, and clinical status after operation were assessed.
Results: In the one-stage surgery group, there were 4 deaths postoperation, and estimated 1, 5, and 15-year survival rates were 96.8%, 93.5%, and 88.4%, respectively. In the staged surgery group, 26 patients died, including 17 cases in the awaiting period after initial intervention. Estimated 1, 5, and 15-year survival rates of staged group were 89.7%, 88.5%, and 69.3%, without significant difference as comparing to one-stage surgery group ($P > 0.05$). Independent predictors of mortality were severe RV hypoplasia ($p < 0.05$) and lower tricuspid valve Z-score ($p < 0.01$). At latest follow-up, most of patients have well clinical status after definitive repairs in the both groups, while re-operation rate was 18.5% (5/27) in the one-stage surgery group and 17.8% (8/45) in the staged surgery group.
Conclusions: Both of one-stage and staged surgical procedure had acceptable surgical outcomes in this retrospective study. Initial intervention is suitable for neonates or younger patients to promote the growth of the RV, and one-stage definitive repair is a beneficial choice for older patients with PAIVS, in whom the growth potential of the RV is limited.

P1733 - THE RASTELLI PROCEDURE LONG TERM OUTCOME IN 25 YEARS OF EXPERIENCE

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Background: The Rastelli operation is the conventional treatment for patients (p) with transposition of the great arteries (TGA) with ventricular septal defect(VSD) and pulmonary stenosis(PS). Its results, however, are suboptimal.
Objective: To analyze the medium and long-term outcome of the Rastelli surgery at our hospital.
Material and Methods: This study analyzed 63p operated on with the Rastelli procedure at our institution between 1991 and 2016. Median age at surgery was 3.8 years (IQR = 2.7-4.9). The median time of follow-up was 12.5years (IQR = 6.9-16). Two anatomical

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variants were identified: TGA with VSD and PS(31p) and double outlet right ventricle with PS(32p). The VSD was unrelated to the aorta in 20 (32%) and restrictive in 9p (14%).
Results: The survival rate at 5 and 10 years was 97% and it was 94% at 15 years. The 98% of the survivors are in CF I-II with good ventricular function. The right ventricular to pulmonary artery (RV-PA) conduit developed a moderate or severe obstruction in 30p (57,7%) and severe regurgitation in 6p. Fourteen patients (26.9%) acquired left ventricular outflow tract obstruction (LVOTO). Seven of them had a non-committed VSD ($p = 0.01$). Ten patients required reoperation to relieve the LVOTO. In the long-term follow-up, the incidence of arrhythmias was 17% (8p) and the mortality was 3,8%(2p). Freedom from interventional catheterization at 5, 10, 15 and 20 years was 74%, 55%, 35% and 28%, respectively. Freedom from reoperations at 5, 10, 15 and 20 years was 83%, 46%, 26% and 19%, respectively. The main indication of reintervention (interventional catheterization = 84% and reoperation = 76%) was RV-PA conduit obstruction and/or incompetence.
Conclusions: - The Rastelli procedure has shown a good long-term survival.

- The most frequent adverse event was RV –PA dysfunction.
- Reinterventions were required in 61% of patients.
- The LVOTO was more frequent in patients with non-committed VSD.

P1741 - AORTIC ARCH RECONSTRUCTION USING AUTOLOGOUS PULMONARY ARTERY TISSUE IN INTERRUPTED AORTIC ARCH REPAIR AS A NEW SURGICAL APPROACH

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Background: This retrospective analysis aims to evaluate our experience using autologous pulmonary artery tissue in patients with interrupted aortic arch (IAA) as a new surgical approach.
Methods: Between 2005-2016 ten patients with the diagnosis of IAA (Type A: 3, Type B:7) and ventricular septal defect (VSD) were operated using autologous pulmonary artery tissue for aortic reconstruction. Six patients were treated initially with hybrid stage 1 approach due to the borderline left-sided heart structures. As a further operation they underwent to a total correction including IAA repair and VSD closure with a combination of bilateral pulmonary artery reconstruction and removal of the ductal stent. The remaining four patients underwent to a primary complete repair. In all patients arch reconstruction was performed under moderate hypotermic (28-30 °C) cardiopulmonary by-pass combined with selective cerebral perfusion. While reconstructing the aortic arch, a quadrangular shaped autologous pulmonary artery tissue was harvested from the anterior wall of the pulmonary artery. The harvested pulmonary autograft was then used to reconstruct the posterior wall of the aortic arch between the aorta ascendes and the aorta descendes (Video Presentation). Anterior wall of the aortic arch was reconstructed with autologous pericardium. The created defect in the main pulmonary artery was replaced with xenopericardium or with autologous pericardium. Nine out of ten patients could be biventricular treated.
Results: Median follow-up time was 70 months. In-hospital mortality occurred in one patient. Freedom from re-intervention and reoperation for aortic arch was %100. There were no hypertension and no bronchial compression observed.
Conclusion: In some cases, especially in IAA type B, applying the direct anastomosis is sometimes not possible or could be dangerous for restenosis. In this condition posterior wall reconstruction with autologous pulmonary artery tissue can be a good alternative to other techniques due to the growth potential of the pulmonary artery and anatomical correction of the arch.

P1757 - INNOVATIVE MIRROR IMAGE TRANSATRIAL TECHNIQUES FOR BIVENTRICULAR REPAIRS IN SITUS INVERSUS AND DEXTROCARDIA

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Background: Correction of congenital heart defects with situs inversus and dextrocardia represent technical challenges due to possible injury to the conduction system. Published approach is trans-ventricular for accomplishing biventricular repair (BVR), with chief surgeon operating from right side of the patient. We present our experience of BVR in these subsets from left side of the patient using innovative pre-operative mirror image techniques.
Materials and Methods: 15 patients with these subsets underwent BVR through innovative trans-atrial approach from left side of the patient. Orientation of intra-cardiac anatomy was obtained pre-operatively by rendering standard illustrations in a reversed and inverted way using standard software. Male: female ratio was 7:8. Mean age and weight were 4 years (4.5 months-31 years) and 10.16 kg (3.4-59 kg) respectively. The surgical spectrum included closure of ventricular septal defects (VSD) (n = 6), complex intra-ventricular tunneling for double outlet right ventricle (n = 3), tetralogy of Fallot (TOF) repair (n = 6), with trans-annular patch (n = 2), with conduit (n = 2), and with double barrel technique (n = 1). One patient underwent Double switch operation (DSO) with Hemi-Mustard procedure for corrected transposition, VSD and pulmonary stenosis. Septal and anterior leaflet detachment was performed whenever necessary (n = 5).
Results: Mean bypass and clamp time was 153 mins and 101 mins respectively. None developed complete heart block. Mean hospital stay was 12.4 days with no hospital mortality. On follow-up, all patients remain in NYHA class I. One adult awaits device closure for a small residual VSD. One required right pulmonary artery plasty (Balloon) after total correction. Patient with DSO got readmitted for supra-ventricular tachycardia and is on beta-blockers. She remains symptom-free till date.
Conclusion: Trans-atrial BVR of these complex subsets is feasible and reproducible. The innovative illustrations facilitated in accomplishing successful repairs while avoiding heart blocks and ventriculotomy. Short-term results are satisfactory.

P1760 - MODIFICATION OF NORWOOD TYPE PROCEDURE AFTER BILATERAL PULMONARY ARTERY BANDING AND DUCTAL STENTING FOR HYPOPLASTIC LEFT HEART SYNDROME

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Bilateral pulmonary banding and ductal stenting can be used as a hybrid alternative to Norwood-type procedure for palliation of neonates. We reported modification of arch reconstruction using stented ductal tissue for hypoplastic left heart syndrome. A two-month-old boy with hypoplastic left heart syndrome underwent bilateral pulmonary artery banding on Day 5 and subsequent ductal stenting. Cardiopulomnary bypass was established through a aortic cannula was inserted in graft which anastomosed with the innominate artery and right atrium was cannulated. The aortic arch between the innominate artery and the left common carotid artery was clamped and the left common carotid artery and the left subclavian artery were snugged with small tourniquets. The main pulmonary artery was transected just proximal to the bifurcation. The origin of the branch pulmonary arteries was resected as a single large button that meant the anterior wall of the main pulmonary artery was completely preserved for the aortic reconstruction. The continuity of the branch pulmonary arteries was restored by folding the pulmonary button. The clamp on the proximal aortic arch was moved to the origin of the innominate artery. The ascending aorta was transected at the level of the proximal pulmonary stump, and then blood cardioplegia was delivered. The area of apposition between the small ascending aorta with the lesser curvature of the aortic arch and the stented ductus was divided towards the orifice of the aortic isthmus. The stented ductal tissue was used as a patch. The aortic arch was reconstructed by sewing the free edges of the divided duct to the opened native aortic arch. Continuity was then restored with the main pulmonary artery and proximal aorta.The branch pulmonary artery bands were removed, and the branches were dilated. The artificial connection between right ventricle and pulmonary artery was created with a 5.0mm EPTFE tube graft.

P1774 - FACTORS ASSOCIATED WITH INTERSTAGE MORTALITY FOLLOWING FIRST STAGE SINGLE VENTRICLE PALLIATION

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Background: Several advances have led to improved hospital survival following first-stage single ventricle palliation. Nonetheless, a number of patients continue to suffer from interstage mortality prior to subsequent second-stage palliation. We aim to study patients' characteristics, anatomic and surgical details associated with interstage mortality in the current era.
Methods: 453 neonates survived to hospital discharge following first-stage palliation. Competing-risks analysis modeled events after first-stage surgery (Glenn, transplantation or death) and examined variables associated with interstage mortality.
Results: Competing-risks analysis showed that 1 year following first-stage palliation, 10% of patients had died, 86% had progressed to second-stage palliation, 1% had received heart transplantation and 3% were alive without subsequent surgery. On multivariable analysis, factors associated with interstage mortality risk were: weight ≤2.5 kg (HR = 2.4(1.2-4.6), p = 0.013), premature birth ≤36 weeks gestation (HR = 2.0(1.0-4.0), p = 0.05), genetic syndromes (HR = 3.2(1.7-6.1), p < 0.001), unplanned cardiac reoperation (HR = 2.1(1.0-4.4), p = 0.05) and prolonged intensive care unit stay >30 days following first-stage palliation (HR = 2.5 (1.4-4.5), p < 0.001). First-stage palliation surgery type (shunt vs. Norwood vs. band) was not associated with interstage mortality except for Norwood with shunt as opposed to RVPA conduit (HR = 5.4(1.5-19.2) p = 0.01). Of interest, while underlying cardiac anatomy was associated with hospital mortality, that was not associated with interstage mortality among hospital survivors (HR = 1.1(0.6-2.2), p = 0.749). Additionally, ECMO use at first stage palliation was not associated with interstage mortality among hospital survivors (HR = 1.1(0.4-3.8) p = 0.800).
Conclusion: In the current era, interstage mortality following first-stage palliation occurs in 10% of hospital survivors. Contrary to hospital death, underlying single ventricle anomaly was not associated with interstage death. Conversely, several patient factors (such as prematurity, low weight and genetic syndromes), clinical factors (the need for unplanned reoperation and prolonged intensive care unit stay following first-stage palliation) are risk factors for interstage mortality. Vigilant post-discharge home monitoring might be warranted to improve survival in those high-risk patients.

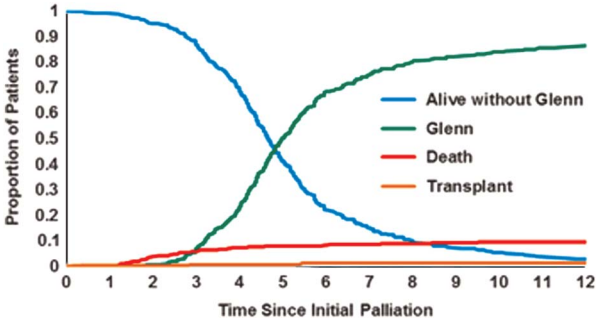

Figure.

P1775 - SIMPLE TECHNIQUE TO PREVENT SUPRAVALVAR PULMONARY ARTERY STENOSIS IN ARTERIAL SWITCH OPERATION

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Objectives: A number of techniques aimed to avoid stenosis after pulmonary artery (PA) reconstruction in arterial switch operation (ASO) have been described. However, this complication is still observed in 4-6% cases. We present our follow-up results of PA reconstruction using single non-bifurcating pericardial patch.
Methods: 87 consecutive patients who underwent ASO for transposition of the great arteries (TGA) between December 2008 and December 2014 were included in this prospective cohort study. Median age and weight at the time of surgery were 12 (3–486) days and 3.6 (2.5–10.3) kg, respectively. Median follow-up duration was 3.2 (0.2–7.3) years. Proximal main PA was routinely reconstructed during aortic cross-clamp period with a single trapezium-shaped, non-bifurcating fresh autologous pericardium patch. Posterior commissure of the neo-pulmonary valve was attached to the pericardium patch using a single stitch. The anastomosis was fashioned during rewarming using continuous 7/0 polypropylene suture.
Results: Freedom from supravalvar pulmonary stenosis defined as Doppler-derived peak systolic pressure gradient (PG) more than 20 mm Hg, and neo-pulmonary regurgitation graded higher than mild, at 7,3 years follow-up was 100%. No patient developed subvalvar right ventricular outflow tract obstruction. Main PA z-score was 0.261,25, neo-pulmonary valve z-score was 0.251,12, left and right PA z-scores were -0.251.03 and -0.551.14,

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respectively. Median peak systolic pressure gradient on the left branch PA was 9 mm Hg (3-28) and on the right 11 mm Hg (4-50). One patient (1.1%) who required two-staged repair for TGA with multiple VSDs had developed clinically significant right branch PA stenosis (PG 50 mm Hg).
Conclusions: Medium-term results of the ASO with application of a single non-bifurcating fresh autologous pericardium patch during reconstruction of main PA demonstrated almost complete freedom from supravalvar main PA stenosis and re-intervention for any right-sided obstructive lesions in all patients.

P1782 - INTRA AND POSTOPERATIVE PREDICTORS OF EXTRACORPOREAL CIRCULATORY SUPPORT REQUIREMENT IN HIGHER RISK CONGENITAL CARDIAC SURGERY

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Background/Hypothesis: Prediction of postoperative low cardiac output and requirement of extracorporeal membrane oxygenation (ECMO) allows for preemptive measures.
Materials/Methods: 510 patients (weight 6.44 ± 3.87 kg;age 300.7 ± 402.6days) undergoing cardiopulmonary bypass (CPB) between 2012-2014 (aged ≤3years, RACHS-1 score ≥3) were included and analyzed. Blood gas, inotropic doses and transfusion were collected at 3 time points (termination of CPB [T1], end of procedure [T2], 2-4 h post ICU admission [T3]). Procedure related ECMO was considered any ECMO within 48 h of surgery. Multivariate logistic regression was used to identify predictors.
Results: A total of 21(4.1%) patients were supported by ECMO. Among those with ECMO initiation on ICU, 6(66.7%) underwent ECPR. Total mortality was 6.3%(n = 32). 4(33.3%) of intraoperative ECMO patients, and 3(50.0%) of ECPR patients died. All patients who underwent semi-elective ECMO in ICU survived. Mortality of non-supported patients was 5.1%(n = 25). Predictors of any ECMO included lower milrinone dose at T1 (r = -2.65;p = .0859), higher epinephrine dose at T2 (r = 8.77; p = .0068), worse grade of residual left atrioventricular valve regurgitation (r = 0.48;p = .0150), residual coronary artery lesion (r = 1.50;p = .0063), aortopulmonary shunt placement (r = 1.55; p = .0003), and longer CPB (r = 0.017;p < .0001)(c = .936). Predictors of ECMO in the operating room included longer CPB (r = .0228;p = .0002), aortopulmonary shunt placement (r = -1.71; p = .0029), higher norepinephrine dose at T1 (r = 4.93;p = .0341), and worse grade of ventricular dysfunction (r = 1.08;p = .0064) (c = .99). Predictors of postoperative ECMO included lower milrinone dose at T1 (r = -4.26;p = .0083), higher norepinephrine at T1 (r = 23.9;p = .0002), residual coronary artery lesion (r = 2.66; p = .0009), procedure involving repair of aortic arch/coarctation (r = 2.32;p = .0037), atrial septectomy (r = 2.27,p = .0038), and longer crossclamp time (r = .0249;p = .0001)(c = .96).
Conclusions: Patients needing ECMO postoperatively had high mortality, especially after ECPR. Patients with high inotropic support, long CPB, and residual anatomic or physiologic lesions are at higher risk to need ECMO. Given high mortality associated with ECPR, a low threshold to place patients with those risk factors on ECMO prior to cardiac arrest may improve survival. Development of a predictive model would be helpful in decision-making.

P1796 - RACIAL ETHNIC AND HEALTHCARE DISPARITIES ASSOCIATED WITH PEDIATRIC CARDIAC SURGERY OUTCOMES IN THE UNITED STATES

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Background/Hypothesis: Despite overall improvements in congenital heart disease outcomes, racial and ethnic disparities continue in the United States. The effect of age on these health disparities is unclear.
Materials and Methods: From the 2012 Healthcare Cost and Utilization Project Kids Inpatient Database (KID), we identified 13,130 records with Risk Adjustment in Congenital Heart Surgery complexity score-eligible procedures. We assessed the effect of race and ethnicity, as well as other risk factors on in-hospital mortality and length of stay (LOS) through multivariate logistic and linear regression modeling with survey weights, stratification and clustering.
Results: No significant mortality differences were found among all race and ethnicity groups across each age group. Black neonates had a longer LOS (Estimate = 8.73 days, p = .0034), as did black infants (Estimate = 1.10 days, p = 0.0253), relative to whites. Income quartile was not associated with mortality or increased LOS in any age group, except for neonatal LOS in the first quartile (Estimate = 4.58 days, p = 0.0325), compared to fourth quartile. Medicare/Medicaid insurance was associated with increased odds of neonatal mortality (odds ratio [OR] = 1.51, p = .0055), and with increased LOS in neonates (Estimate = 4.26 days, p = .0009) and infants (Estimate = 1.52 days, p = .0181) relative to private insurance. Chronic conditions and hospital-acquired injuries/complications were strongly associated with mortality and LOS in all age groups.
Conclusions: Racial/ethnic disparities in congenital heart surgical outcomes continue in the United States. The number of chronic conditions and incidence of medical injuries/complications are significant contributors to mortality and LOS.

P1805 - A RARE CONGESTIVE HEART FAILURE REASON IN AN INFANT AORTICO LEFT VENTRICULAR TUNNEL

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The aortico-left ventricular tunnel is a very rare congenital malformation, characterized by an abnormal communication between the ascending aorta and the left ventricle. Generally the tunnel is situated above the right coronary sinus. The pathogenic mechanism of this congenital anomaly is not completely understood yet. In most cases the first symptoms appear in early infancy. Clinical presentation and the hemodynamics are similar to those of aortic valve insufficiency. Definitive diagnosis is established by echocardiography and angiocardiography. Early operation is recommended before the aortic annulus becomes excessively dilated. The method of choice for surgical correction is a patch closure of the aortic orifice of the tunnel. we report a case of aortico-left ventricular tunnel that was associated with right ventricular outflow tract obstruction due to the aneurysmal tunnel. The aortico-left ventricular tunnel was closed so as to treat the gradually increasing left ventricular systolic dysfunction.

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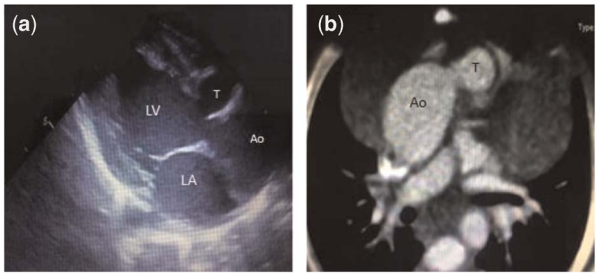

Figure 1.
(a) Transtoracic echocardiography in the parasternal long axis view demonstrating the entire extent of the aortico-left ventricular tunnel and the dilated aortic root, (b) Computed tomography demonstrates the tunnel itself bulged into the RVOT (b). Ao = aorta, T = tunnel, LV= left ventricle, RV = right ventricle, LA = left atrium.

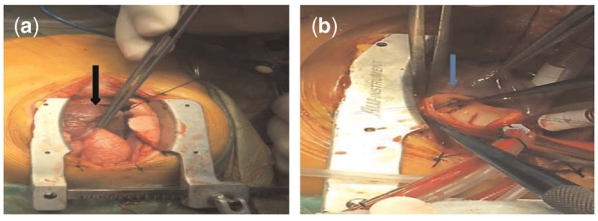

Figure 2.
(a) Operative view of the aortico-left ventricular tunnel, (b) Surgical repair by patch closure of its aortic entrance next to the right coronary cusp.

P1809 - EARLY RESULTS 2 YEARS OF CARDIOCEL® PATCH IMPLANTATION IN CHILDREN

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Hypothesis: Early results after 2 years of CARDIOCEL® patch (Admedus Regen Pty Ltd, Perth, WA, Australia) in paediatrics for congenital heart disease repair shows an unexpected early re-stenosis when sutured in aortic position.
Materials and Methods: Single centre retrospective study of all patients aged 18 year-old and under, operated for congenital heart diseases. Standard follow-up was performed, clinically and with echocardiograms. In case of re-operation for graft failure, the patch was removed and sent to pathology.
Results: Between March 2014 and April 2016, 101 patients with a congenital heart disease had a surgical repair using the CARDIOCEL® patch. The mean age was 7 months old (range: 3 days – 18 years) and the mean weight was 6,7 kg (range: 1,83-96 kg). The mean bypass time was 156 min (+74 min) and the mean aortic cross clamp time was 103 min (+49 min). No infections, no intraoperative implantation difficulties were related to the patch. The mean follow up was 175 days (range: 4-726 days). The 30-day overall post-operative mortality was n = 4 (3,8%), none were related to graft failure. 5 infants were re-operated because of graft failure, all in aortic position. The indication for the patch in aortic position was truncus arteriosus, coarctation and aortic arch hypoplasia repair. The mean time between the first and the second operation for graft failure was 189 days (range: 16-480 days).
Conclusions: Our experience shows a good tolerance of the patch in septal, valvar and pulmonary artery position. However, we experienced graft failures in infants in aortic position.

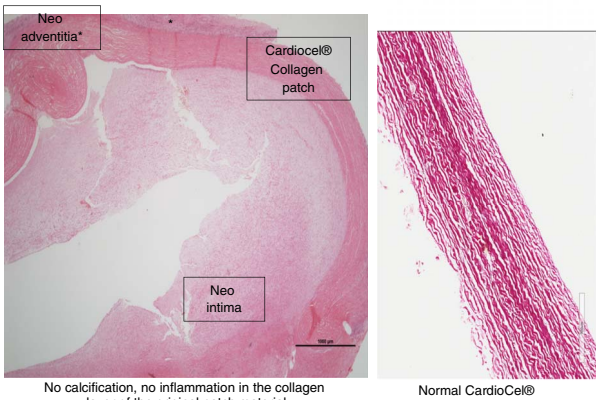

Figure.

P1811 - RAMP UP AN ITERATIVE TOOL TO ENSURE PEDIATRIC HEART PROGRAM PERFORMANCE

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Background: Clinical care of patients with complex congenital heart disease is a multidisciplinary undertaking. Critical human resource changes in a heart program can threaten its ability to deliver consistent outcomes. We describe a "Ramp Up" process, employed three times in a small-volume high-performance program, to mitigate risk delivering pediatric cardiac care.
Methods/Materials: The IWK Heart Program is comprised of pediatric cardiologists (n = 5), cardiac surgeons (n = 2), cardiac anesthetists (n = 1) and intensivists (n = 4). The program provides care to patients of the four Atlantic provinces with all congenital cardiac pathologies and performs all surgeries except ventricular assist device and transplantation. Outcome data for each procedure is submitted to a national database (Congenital Cardiac Surgery Database, CCSDB, Hospital for Sick Children, TO) with caregiver consent. Three times over the past 12 years (2003, 2006, 2015) there have been sufficient disruptions to the staffing complement (retirement or relocation) that we have employed a Ramp Up strategy to ensure program performance (Fig. 1a).
Results: Overall 12y program mortality rates remain low (3.3%)and comparable to national standards (4.3%, Fig. 1b). Mortality in benchmark cases (VSD, ASD secundum, complete AVSD, Fontan, tetralogy of Fallot, coarctation, arterial switch and Stage 1) was likewise comparable to national data (IWK 2.8% v CCSDB 2.8%). We also instituted complication monitoring which shows that we have similar outcomes to STS data.
Conclusions: "Ramp Up" requires buy-in from all local team members. External validation of outcomes is critical to provide an arms-length opinion as to the preparedness of the team to move on to cases of greater complexity based on qualitative and/or quantitative observations. "Ramp Up" has proven invaluable as a concept and practice to ensure delivery of the best possible outcomes in pediatric cardiac surgery.

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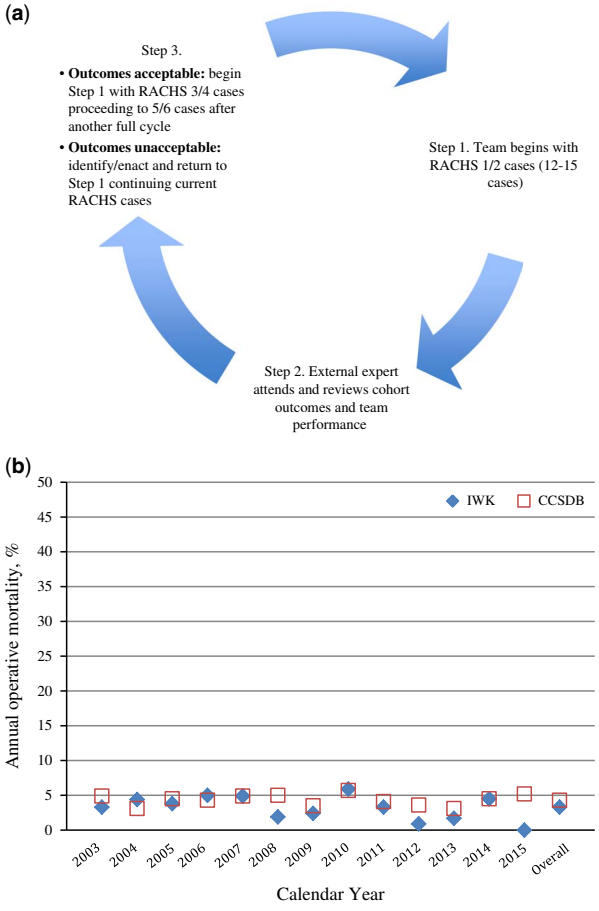

Figure.
(a) Ramp Up process. (b) Annual mortality rates (IWK v CCSDB aggregate 2003-2015) illustrate the maintenance of stable outcomes despite change in surgical staff members (2003, 2006) and anesthesia staff members (2015). (Note annual operative scale is truncated at 50%)

P1812 - QUALITY IMPROVEMENT INITIATIVES TO REDUCE THE WAITING TIME AND MORTALITY FOR PATIENTS UNDERGOING SURGICAL CORRECTION TETRALOGY OF FALLOT IN INSTITUTE JANTUNG NEGARA

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Introduction: The incidence of Congenital Heart Diseases (CHD) among Malaysian children is about 8 to 10 per 1,000 live births which half will require some form of intervention. In IJN the total number of patients undergoing intervention for congenital heart lesions is around 1400 per year.
Objectives: The objective of this study was to reduce the mortality for TOF correction to below 1% as a part of the initiative of Quality Improvement and Patient Safety standards from Join Commission International and to bring down the waiting list for patients with TOF awaiting surgical correction.
Methods: Between January 2015 and December 2016, 230 patients underwent surgical correction of Tetralogy of Fallot. This was the period during which the quality and patient safety initiative was conducted in the institute. The data was obtained from the IQIC redcap database. The patients were screened according to the age, weight complexity of the lesion and distributed accordingly to the surgical teams. Based on the individual surgeons numbers and performance we limited the credentialing the number of surgeons who performed the surgical correction.
Results: The total number of patients on the waiting list reduced from 216 to 22. The median age for patients undergoing total correction reduced from 4.8 years to 2 years. The average slotting of cases per week for TOF correction increased to 10 cases per month. The average mortality of patients who underwent total correction reduced to 0.97%. Based on the performance the number of surgeons who were credentialed to perform the surgery were reduced from 6 to 3 surgeons.
Conclusion: Centralized waiting list and patient selection based on complexity and age and referral to individual surgeons and limiting the credentialing to perform the surgery based on volumes and outcomes has helped to reduce the mortality and waiting times following surgical correction.

P1813 - SUCCESSFUL RE IMPLANTATION OF THE ABNORMAL LEFT CORONARY ARTERY ARISING FROM THE RIGHT VALSALVA SINUS AND COURSING BETWEEN THE ASCENDING AORTA AND THE PULMONARY TRUNK

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Background: Congenital abnormality of the coronary arteries is one of the major causes of sudden death among healthy young people. Although early diagnosis and safe and sure treatment are inevitable to avoid disastrous clinical course, they are sometimes difficult. We present our experience of successful re-implantation of the abnormal left coronary artery (LCA).
Case: A 14-year-old boy without any symptom with non-specific ECG-change was referred to us with a diagnosis of an abnormal LCA originating from the right Valsalva sinus (RVS) and coursing between the ascending aorta and the pulmonary trunk (Fig 1). The origins of the RCA and LCA formed a short common trunk. Cardiac scintigraphy showed no myocardial ischemia. According to the ACC/AHA guideline, surgical operation is indicated with Recommendation Class-1 in this type of coronary abnormality. Some surgical options were considered, such as fenestration, unroofing, CABG, pulmonary translocation and LCA re-implantation. The first two methods were rejected because the LCA had no intramural course. CABG was not suitable because the LCA flow was not reduced and early graft obstruction due to flow competition was concerned. Pulmonary translocation, in our opinion, has little effect to relieve the LCA obstruction. LCA re-implantation is easily indicated when the LCA had a separate ostium, but our case had single ostium. After all, we chose LCA re-implantation. The LCA was divided just distal to the LCA-RCA bifurcation. The LCA was dissected free as distal as possible, over the LCA-LAD bifurcation. The proximal end of the LCA was incised to form a good shape of "Cobra head", which was anastomosed to the left Valsalva sinus. Postoperative course was uneventful and postoperative CT showed non-obstructive LCA (Fig2). In conclusion, re-implantation of the LCA originating from the RVS can be achieved safely even when the LCA and RCA had common trunk.

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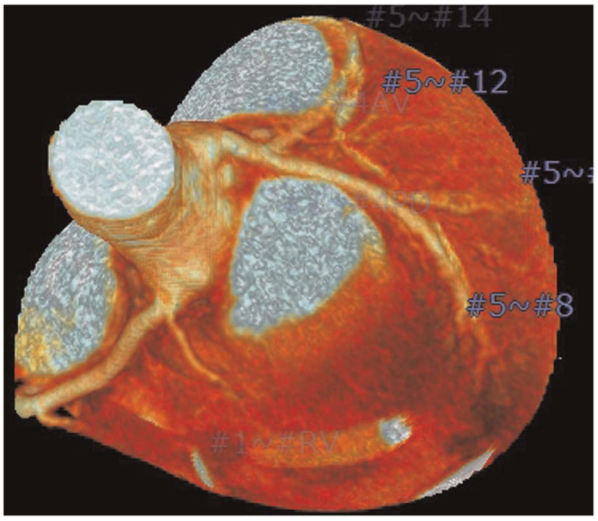

Figure 1.

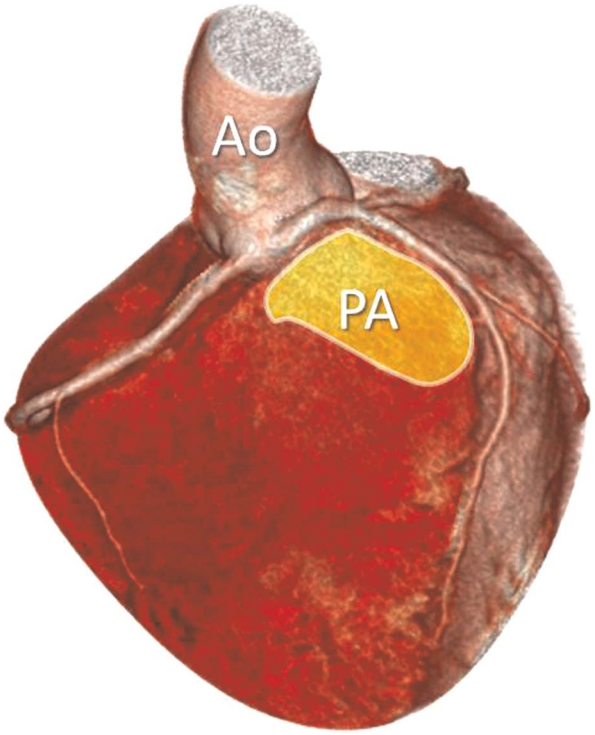

Figure 2.

P1814 - THE INFLUENCE OF ISOLATED BIDIRECTIONAL CAVOPULMONARY CONNECTION ON PULMONARY ARTERY

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Cardiology Reserach Institute, Tomsk Nrmc, Cardiovascular Surgery Department, Tomsk-Russia

Background: Bidirectional cavopulmonary connection (BCPC) is the best interim stage for hemodynamic correction of congenital heart diseases with single ventricle (SV). Still the problem of additional source of pulmonary blood flow at BCPC for adequate flow is unsolved.
Materials and Methods: 89 patients at the age from 2 months to 13 years (Me = 0,64; IQR:0,5-1,18) with SV were included into the study. They were not performed heart catheterization at the stages of hemodynamic correction with pulmonary angiography to assess achitoctonics of pulmonary blood flow and index of pulmonary artery (PA): Nakata index, McGoon ratio and Reddy index from 2012 to 2014 years.
Results: Initially, Nakata index was Me = 326,0 мм2/м2 (IQR:241,5-410,0), Reddy index - Me = 215,0 мм2/м2 (IQR:151,5-283,5) and McGoon ratio Me = 1,8 (IQR:1,7-2,2). In 30 months after BCPC increase of the following PA indices was marked: Nakata index до Me = 336,5 мм2/м2 (IQR:240,0-415,5), p = 0,717, Reddy index till Me = 222,0 мм2/м2 (IQR:140,5-288,8), p = 0,147 and McGoon correlation increased up to Me = 2,0 (IQR:1,7-2,2), p = 0,288. All PA indices and McGoon correlation after BCPC corresponded to normal values. The comparative analysis of right and left PA diameters showed significant difference, p = 0,005: right PA - Me = 12,05 mm (IQR:10,57-14,4), left PA - ЛА - Me = 11,5 (IQR:9,57-12,6). Diameters of distal PA branches also showed significant difference: right distal PA - Me = 11 mm (IQR:8,35-11,5), left distal PA - Me = 9,5 mm (IQR:8,4-10,8).
Conclusion: BCPC without additional source of pulmonary blood flow provides sufficient pulmonary flow. However, asymmetric growth of PA branches is marked.

P1825 - THE CLINICAL ANALYSIS OF 99 CASES WITH CONGENITAL VASCULARRING

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Bacground and Objevtive: Congenital vascular rings (CVR) are rare. This study is to evaluate the clinical characteristics,diagnosis and prognosis of CVR in children.
Methods: 99 cases of CVR in our hospital from January 2006 and December 2015 were retrospective riviewed.According to the prognosis of divided into survival group and death or give up the treatment group, the risk factors of poor prognosis analysis.
Results: 99 cases with congenital vascular rings included 55 males and 44 females,age median 7.2 months.The diagnosis including pulmonary artery sling in 51 cases,double aorta arch in 32 cases, right aortic arch with left ductus arteriosus in 15 cases,left aoaic arch with vagus right subclavian artery in 1 case.Merge heart malformations in 64 cases (64.6%), of which the merger pure ASD in 31 cases, VSD in 2 cases, 9 cases with ASD + VSDand 22 cases of complex cardiac anomalies. Tracheobronchial stenosis in 69 cases, esophageal stenosis in 14 cases, 10 cases of laryngeal cartilage dysplasia, 9 cases of congenital bronchial pulmonary hypoplasia. The diagnosis accuracy of echocardiographic was 57.4% and that of computed tomography angiography was 91.6%,26 cases checks on the fiber bronchoscope were found airway stenosis; 14/17 (82.3%) line barium esophageal imaging examination in esophageal pressure distortion and narrow. 57 cases of congenital vascular ring completed the operation,including 4 cases died, postoperative offline difficult to give up treatment of 6 cases.Non-surgical treatment of 42 cases,among which 1 case death and 10 cases of offline difficult to give up treatment. Dead or abandoned children, age, and proportion of the pulmonary artery sling with pulmonary dysplasia group is significantly higher than survival.

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Conclusion: Eehocardiography combined with CTA is the best way to diagnose vascular ring.Type of vascular ring,age and the combining pulmonary hypoplasia were risk factors worsening the patients'prognosis.

P1833 - A RETROSPECTIVE STUDY OF VARIOUS TECHNIQUES OF SURGICAL CLOSURE OF ATRIAL SEPTAL DEFECT

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Devadoss Multispeciality Hospital, Paediatric Cardiac Surgery, Madurai-India

Hypothesis: A Retrospective study of Various surgical techniques used for ASD closure by a single surgeon form June 2013 TO December 2016 and its merits and demerits.
Material and Methods: All cases of surgical closure of ASD was evalvauated restrospectively from June 2013 to December 2016 by a single surgeon. There was 193 cases of Surgical closure of ASD of various age form 3 years to 48 years. All cases were not suitable for ASD Device closure and reffered for surgery by Paediatric cardiologist. Out of 193 cases, 136 cases were done by standard Sternotomy,44 cases by Lower partial sternotomy and 13 cases by Right Anterolateral Thorocotomy.
Results: All cases do have similar CPB and Cross clamp time, irrespective of Technique uses. Only 11 of the 193 cases were ventilated for more than 4 hours post operatively in the ICU. All the 44 Lower partial sternotomy cases were less than 10 years of age and they were dischared on the 3rd Post operative day. No difference in Hospital stay between the Standard sternotomy and Anterolateral thorocotomy cases.
Conclusion: Lower partial sternotmy is an effective and surgically safe alternative to ASD device closure in children less than 10 years and less than 25 kg weight, compared to standard Sternotomy or Anterolaterlal thorocotomy surgical ASD closure techniques.

P1835 - HYBRID APPROACH IN PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND COLLATERALS COIL OCCLUSION AND RIGHT VENTRICLE PULMONARY ARTERIES CONDUIT IMPLANTATION WITH POST REPAIR ANGIOGRAPHY IN A HYBRID OPERATING ROOM. INITIAL EXPERIENCE

Marcelo Jatene[1], Leonardo Miana[1], Leandro Batisti[1], Raul Arrieta[2], Carlos Pedra[3], Germana Coimbra[2], Marcelo Ribeiro[3], Rodrigo Costa[3], Ieda Jatene[4], Fábio Jatene[5]
Incor HC FMUSP and HCOR, Pediatric Cardiovascular Surgery, São Paulo-Brazil[1]; Incor HC FMUSP, Pediatric Cardiovascular Surgery, São Paulo-Brazil[2]; Hcor, Pediatric Cardiovascular Surgery, São Paulo-Brazil[3]; Hcor, Pediatric Cardiology, São Paulo-Brazil[4]; Incor Hc Fmusp And Hcor, Cardiac Surgery, São Paulo-Brazil[5]

Introduction: Pulmonary Atresia with Ventricular Septal Defect and major aortopulmonary collateral arteries surgical timing and approach is a challenging situation. MAPCAS surgical intervention is difficult and time consuming. Sometimes various surgical approaches are necessary. Hybrid procedures bring the best of two interventions in a lot of congenital cardiac diseases.
Objective: We sought to evaluate an initial experience with an hybrid (interventional + surgical) approach in patients with Pulmonary Atresia with Ventricular Septal Defect and major aortopulmonary collateral arteries. Methods: Retrospective study. Between November 2014 and November 2016, 18 consecutive patients with median age of 10.5 months and median weight of 7.5 kg.
Results: Thirteen out of 18 patients (72.2%) were submitted to sucessfull coil collateral embolization pre-bypass. Four unifocalizations and three PA enlargements were performed. VSD closure were perfomed in 14 patients (77,7%), in the other 4 patients, RV-PA conduits were implanted and VSD were left open due to poor pulmonary vascular bed. Bypass and cross-clamp median time was 156.5 and 83 minutes, respectively. Ten patients (55.5%) were submitted to post-repair angiography through RV cannulation. In five (50%) pulmonary angioplasty and stent implantation were performed in the same operative time. Intubation and ICU median time was 4.5 and 9 days, respectively. The 30-day mortality was 11.1% (2 patients) and there was one late death.
Conclusion: Hybrid OR allows a combined and integrated approach to Pulmonary Atresia with Ventricular Septal Defect and major aortopulmonary collateral arteries patients enabling a thorough procedure in one surgical time.

P1845 - MITRAL VALVE REPLACEMENT USING STENTED BOVINE JUGULAR VEIN GRAFT (MELODY VALVE) IN INFANTS AND SMALL CHILDREN

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Background: Melody® valve (Medtronic, MN) implantation in the mitral position is an emerging alternative to a mechanical valve especially in younger children. It is implantable in a small mitral valve (MV) annulus and does not require long-term anticoagulation. The aim of the current study was to evaluate our initial experience with mitral valve replacement (MVR) with Melody® in small children.
Method: The clinical records of patients who underwent MVR using Melody® from 2014 to 2016 were retrospectively reviewed. The Melody® (22 mm) valve was prepared by sewing a 3.5 mm Gore-Tex tube graft at the middle of the stent as a cuff and resecting one or three zigs to avoid left ventricular outflow tract (LVOT) obstruction. All values are represented as median with range.
Results: Five patients (age, 11 months, 5-16 months; weight, 6.8 kg, 4.6-8.6 kg) were included. All patient had a dysplastic MV with severe stenosis (n = 3) and/or regurgitation (n = 3). Three patients underwent Melody® valve MVR as a salvage procedure after mechanical valve thrombosis (n = 2) or tissue valve dysfunction (n = 1). Cardiopulmonary bypass time was 160 minutes (60–199 minutes) and cross-clamp tim e was 106 minutes (40-134 minutes). The valve was dilated to 18 mm (n = 2) or 20 mm (n = 3). Intraoperative echocardiography revealed no or trivial regurgitation in all patients with a mean pressure Doppler gradient of 2 mmHg (1–4 mmHg), and peak LVOT gradient of 5 mmHg (0–12 mmHg). A patient required intraoperative revision for LVOT obstruction due to the stent. All the patients but one were discharged home. There was one patient who had sudden death at 3 months after surgery.
Conclusions: Melody® valve MVR is a viable alternative to mechanical MVR in small children not only as a primary strategy but as a salvage strategy for dysfunctional valves. The long-term outcome remains to be investigated.

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P1853 - PULMONARY ARTERY BANDING AS A PALLIATION; EARLY AND MID TERM RESULTS

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Mehmet Akif Inan Educational and Research Hospital, Pediatric Cardiac Surgery, Sanliurfa-Turkey[1] Kartal Koşuyolu Educational and Research Hospital, Pediatric Cardiac Surgery, Istanbul-Turkey[2] Kartal Koşuyolu Educational and Research Hospital, Clinic of Anesthesiology, Istanbul-Turkey[3] Marmara University, Marmara Faculty of Medicine, Department of Public Health, Istanbul-Turkey[4]

Pulmonary Artery Banding (PAB) is the first palliative procedure of a staged approach to complex congenital cardiac anomalies in patients with unrestricted pulmonary blood flow (PBF).
Materials and Methods: The medical records and follow-ups of 81 patients who underwent PAB between January 2011 and June 2016 were reviewed. The study subjects were grouped into 2 categories based on their cardiac diagnoses. Group 1 (n = 26, 10 female, 16 male) included single-ventricle variants with unrestricted PBF and no evidence of systemic outflow tract obstruction. Group 2 (n = 55, 32 female, 23 male) involved candidates for terminal biventricular repair and critical medical conditions with unrestricted PBF. The endpoints were assigned as: early death after PAB, death following stages, total correction for biventricular physiology or definitive operation for single-ventricle variants, awaiting definitive operation and declined follow-up.
Results: A total of 81 patients (39 men, 42 women; median age 4 months, IQR:6) were identified. PAB mortality at the first stage was 12.3%, and was markedly elevated in Group 2 ($p < 0.01$) but not related to the gender, age, hospitalization or duration of PAB. The band was inadequate in 3 patients at the follow-up. Median duration of PAB (7 months in Group 1, IQR:10, 27.5 months in Group 2, IQR:5) was not related to the age, diagnostic categories and overall results. The overall mortality during the period studied was 18.4% and significantly higher in Group 2 than Group 1 ($p < 0.01$). Thirty-five patients (44.4%) have had a final definitive repair, and 24 (29.7%) patients are awaiting their final definitive repair. 6 patients (7.4%) were declined follow-up.
Conclusions: PAB is indicated in both patients with excessive PBF and single ventricle variants and patients with two balanced ventricles that are not amenable to early primary repair. However the role of palliative PAB in biventricular repair remains controversial with high early mortality rate.

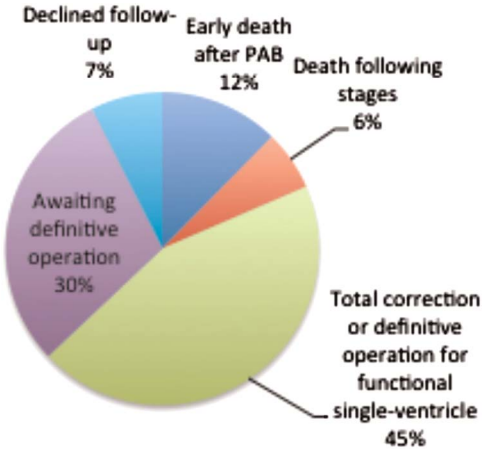

Figure 1.
Outcome of patients after PAB at follow-up.

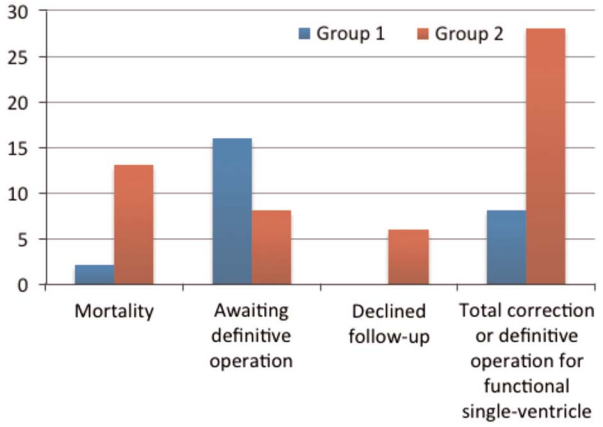

Figure 2.
The distribution of endpoints after PAB procedure based on their cardiac diagnoses. Group 1 includes candidates for single-ventricle variants with unrestricted PBF and no evidence of systemic outflow tract obstruction. Group 2 includes candidates for terminal biventricular repair and critical medical conditions with unrestricted PBF.

P1856 - MID TERM OUTCOMES OF ARTERIAL SWITCH OPERATION A SINGLE CENTRE EXPERIENCE

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Background: The arterial switch operation (ASO) is the surgical approach of choice for d-transposition of the great arteries (d-TGA). Our study evaluates the midterm outcomes in this population.
Materials and Methods: In this retrospective study 78 such patients between January 2011 and July 2016 were included. The mean age was 21 ± 143 days.68%(53) were male and 32%(25) were female. 64%(49) had an intact septum, 46%(29) had a ventricular septal defect, and 12%(10) had Taussig Bing anomaly. The usual coronary pattern (Yacoub type A) was found in 76% (60) abnormal coronary pattern was found in (types B, C, D, E) 24% (18). Palliation prior to arterial switch was performed in the form of balloon atrial septostomy in 8% (7) and maintenance of ductal patency by stenting 10% (8) and prostaglandin infusion 15% (12) of the patients. The follow-up period was 260 patient-years.
Results: The all-cause mortality rate was 7.5% in the early postoperative period. The 10-year rates of freedom from significant neo aortic regurgitation, arrhythmias, pulmonary stenosis were 94.6 ± 4.6%, 97.8 ± 2.3%, 95 ± 3.4% respectively. Most of the patients (98%) were functionally at New York Heart Association (NYHA) class 1 activity. By multiple logistic regression, the following variables were significant risk factors for early mortality: dynamic left ventricular outflow tract obstruction (LVOTO) resection at time of ASO ($p < 0.001$; odds ratio [OR] 19.0; 95% confidence interval [CI] 3.3 to 111.1), a weight <2.5 kg at time of ASO ($p < 0.001$; OR 22.5; 95% CI 5.5 to 91.4), presence of a severe pulmonary artery hypertension ($p < 0.001$).
Conclusion: ASO remains the primary procedure of choice for simple TGA and acceptable choice for complex TGA in terms of overall survival and freedom from re intervention. Long term follow up is essential for further results.

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P1874 - PREOPERATIVE ADMINISTRATION OF SILDENAFIL TO PEDIATRIC PATIENTS WITH CONGENITAL CARDIAC SHUNTS AND MODERATELY ELEVATED PULMONARY VASCULAR RESISTANCE

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Background and Objective: Preoperative administration of pulmonary vasodilators has been considered in congenital heart disease with pulmonary hypertension. One could use this strategy to analyse characteristics of pulmonary circulation rather than to define operability, which involves several other factors. We therefore decided to test for pulmonary vasoreactivity by administering sildenafil preoperatively to pediatric patients. We also measured serum levels of inflammatory mediators.
Patients and Methods: Patients (n = 29) were aged 11.8 [7.5–20.8] months (median and interquartile range), all of them with unrestrictive cardiac communications. At cardiac catheterization, pulmonary vascular resistance (PVR) was 4.7 [3.7–7.9] units x m2. Oral sildenafil (1.0–5.0 mg/Kg/day) was administered for 8 – 70 days until development of pulmonary congestion (desired effect) or confirmation of "no effect" (minimum, 60 days). Sildenafil effect was further tested by measuring pulmonary-to-systemic blood flow ratio (Qp/Qs, echocardiography). Interleukin 6 (IL-6) and intercellular adhesion molecule 1 (ICAM-1) were measured by chemiluminescence before and during sildenafil administration.
Results: At cardiac catheterization, a 33% decrease in PVR was observed during nitric oxide inhalation ($p < 0.001$). Subsequently, sildenafil administration was followed by a 10% increase in Qp/Qs ratio (2.0 [1.4–2.2] to 2.2 [1.7–2.5], $p = 0.017$) associated with a 2% increase in peripheral oxygen saturation (93% [90–96%] to 95% [93–97%], ($p = 0.022$). Thus, 71% of patients responded to inhaled nitric oxide, while 65% of cases responded to sildenafil. However, there was no correlation between responses ($r = 0.12$, $p = 0.564$). Sildenafil administration resulted in a 46% decrease in circulating IL-6 ($p = 0.027$); in responders, there was a 21% decrease in ICAM-1 ($p = 0.011$). Twenty-six patients were subsequently considered as suitable for surgery.
Conclusions: Patients responded differently to acute inhalation of nitric oxide and sustained use of oral sildenafil. Both responses may be clinically relevant. Changes in mediators of inflammation suggest effects of sildenafil administration beyond simple pulmonary vasodilation.

P1883 - PEDIATRIC VENTRICULAR ASSIST DEVICE EXPLANTATION PREDICTORS OF SUCCESSFUL WEANING

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Background/Hypothesis: Long term pediatric mechanical support as a bridge to heart transplantation is associated with significant risks of pump thrombosis and stroke. Ventricular assist device (VAD) weaning protocols for children are not standardized. We evaluated three children that were successfully separated from VAD support following life threatening thromboembolic complications in order to identify predictors of successful weaning.
Materials and Methods: A single center retrospective review was performed of all VADs (22 devices, 21 patients) implanted from April 2008 to December 2016. Of 4 explants performed, 3 were in children that were analyzed for hemodynamic, biochemical and echocardiographic parameters to gain insight into why explantation was successful. The fourth explant was excluded as it was a temporary RVAD post heart transplant. Echocardiographic data pre-VAD implantation was compared to data obtained either following explant (emergency explantation, N = 2), or with the device turned off in the setting of a pump change (N = 1). Data was analyzed using paired student t-tests.
Results: Demographic data and devices are listed in Table 1. The 3 patients with LVADs were explanted due to thromboembolic complications. Mean duration of VAD support for the 3 patients was 37 days. Children explanted demonstrated significantly reduced LV end diastolic volume (LVEDV), LV end diastolic dimension (LVIDd) and increased LV ejection fraction (EF; Table 2). Brain natriuretic peptide (BNP) levels were reduced peri-VAD explantation. All three patients are alive and well at 3 years follow-up with one patient undergoing heart transplant.
Conclusions: Children undergoing VAD implantation have a very high risk of thromboembolic complications. Significant LV functional recovery was observed in patients explanted. We recommend that pediatric VAD recipients should have a predetermined, structured weaning protocol and those with significant reduction in LV size, improvement in LVEF, and decreased BNP should be considered for explantation..

Table 1.

N = 21	PATHOLOGIC PROCESS	Devices Implanted	LVAD	BIVAD	RVAD
Dilated Cardiomyopathy	14	BERLIN	8	2	
Single Ventricle Failure	3	PEDIMAG	1		
Restrictive Cardiomyopathy	2	HEARTWARE	6	2	
Anomalous Coronaries	2	CENTRIMAG	2		1

Table 2.

N = 3	Average pre-VAD Z-score	Average peri-explant Z-score	Δ pre implant to peri-explant Z-score	P value
Echo variable (z-score)				
LVEDV	9.4 ± 5.7	2.5 ± 3.4	-6.9 ± 2.9	0.03
LVIDd	7.8 ± 2.8	2.9 ± 2.3	-4.9 ± 1.9	0.02
LVEF	-8.3 ± 1.7	-3.9 ± 3.3	+ 4.4 ± 2.4	0.05
BIOMARKERS	Average pre-VAD value	Average peri-explant value	Aperi-implant to explant	P value
BNP	2120 ± 1225	130 ± 86.23	-1989 ± 1302	0.06

P1893 - USE OF BIOPULMONIC® CONDUIT FOR RVOT RECONSTRUCTION SINGLE CENTRE MID TERM OUTCOMES

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Introduction: We present mid-term outcomes of the Biopulmonic® (BioIntegral Surgical Ldt.) conduit for RVOT reconstruction.
Methods: All patients undergoing RVOT reconstruction between January 2013 and August 2016 at Southampton University Hospital were retrospectively reviewed.
Results: Thirty-seven conduits were used in 35 patients. Median age at implant was 3.7 years (2 months - 29.9 years). Diagnoses included Tetralogy of Fallot spectrum (52,%), aortic valve disease (10,8%), transposition of great arteries (10,8%), pulmonary atresia with intact ventricular septum (5,4%), double outlet right ventricle (5,4%), interrupted aortic arch (5,4%), truncus arteriosus (5,4%), congenital corrected transposition of great arteries (2,7%). Surgical indications included pulmonary valve anomalies (48.6%), completion of repair after shunts (16%), pulmonary arteries stenosis (13,5%), Ross procedure (5,4%), conduit compression (5,4%), endocarditis (5,4%), coronary stenosis (2,7%) and tricuspid atresia (2,7%). Preoperative transpulmonary gradient was 49 mmHg (3.1–110 mmHg). Median bypass time was 194 min (89–364 min) and median aortic cross-clamp time of 112 min (0–224 min). Thirty patients (92%) underwent redo surgeries, and a previous RV to PA conduit was replaced in 28 procedures (76%). Additional procedures were performed in 31 patients (83,7%). At discharge, median peak gradient across the pulmonary valve was 11.29 mmHg (1,70–29.4) and pulmonary regurgitation was absent or trivial in 65%. At latest follow-up (mean 269 days) gradient rose to 32.5 mmHg (8.76–112) in 22 patients (59%), and two-thirds patients had no or trivial pulmonary regurgitation. There were no conduit-related deaths, but two patients required early BioPulmonic® conduit replacement and one underwent early pulmonary artery angioplasty.
Conclusion: The BioPulmonic® conduit is safe and provides very good hemodynamic parameters at mid-term. Further data is needed to evaluate its long-term performance.

P1895 - A NOVEL BIOABSORBABLE PULMONARY VALVED CONDUIT – SURGICAL TECHNIQUE AND INITIAL CLINICAL RESULTS

Zsolt Prodan[1], Tomasz Mroczek[2], Mohd Azhari[3], Zsolt Nagy[1], Janusz Skalski[2], Sivakumar Sivalingam[3], Thierry Carrel[4], Martijn Cox[5], Oleg Svanidze[6]
Children Heart Center, Pediatric Cardiac Surgery, Budapest-Hungary[1]; Jagiellonian University, Pediatric Cardiac Surgery, Krakow-Poland[2]; National Heart Institute, Pediatric Cardiac Surgery, Kuala Lumpur-Malaysia[3]; University Hospital, Pediatric Cardiac Surgery, Bern-Switzerland[4]; Xeltis Bv, Research And Development, Eindhoven-The Netherlands[5]; Xeltis Ag, Medical Affairs, Zurich-Switzerland[6]

Background/Hypothesis: The bioabsorbable pulmonary valved (PV) conduit is designed to allow patient's own cells to infiltrate and replace the polymeric device material by a process called Endogenous Tissue Restoration, leading to natural tissue growth. After implantation the conduit while remaining fully functional is gradually being replaced by the components of native tissue. This prospective feasibility study evaluated safety and performance of a novel bioabsorbable PV conduit in pediatric patients requiring right ventricular outflow tract (RVOT) reconstruction.
Materials and Methods: Twelve patients aged 2–12 years (50% males) requiring RVOT reconstruction have been enrolled in the study from July to December 2016. In 6 patients various conduits were previously implanted (3 Contegras, 2 Hancocks and 1 homograft), in 3 patients transannular patch RVOT repair (including 1 with monocusp valve) was previously performed. In 3 patients RVOT has been intact. The bioabsorbable PV conduits (16mm -5, 18mm – 7) have been implanted using monofilament continuous sutures (6-0 for distal, 5-0 for proximal anastomoses). The device performance evaluation has been performed by transthoracic ultrasound.
Results: All 12 patients have been successfully recovered from the procedure and completed 6 weeks to 6 months follow-up. In all cases successful implantation has been achieved. No device related complications were reported to date. Ultrasound studies have demonstrated anatomical (conduit diameter, leaflet and wall thickness) and functional (hemodynamics, leaflet motion, absence of thrombosis) stability of the valved conduits in all patients at up to 6 months postoperatively.
Conclusions: The bioabsorbable PV conduit demonstrated ability to be successfully implanted in various anatomical conditions using standard surgical techniques. The early clinical outcomes are looking promising for this technology with the potential to improve results of RVOT reconstruction. Long term follow-up studies to confirm these observations are warranted. ((Figure 363)).

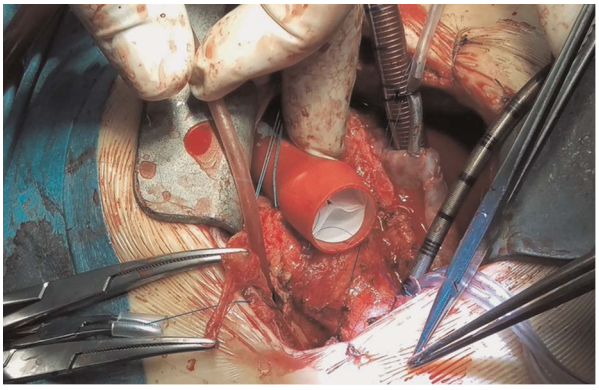

Figure.

P1898 - ANOMALOUS LEFT CORONARY ARTERY FROM "RIGHT" PULMONARY ARTERY; RARE BUT DISTINCTIVE ANOMALY OF DIAGNOSTIC AND SURGICAL IMPORTANCE

Masaaki Kawada[1], Ko Yoshizumi[1], Koichi Kataoka[2], Kensuke Oka[2], Daisuke Matsubara[2], Yu Suzuki[2]
Jichi Children's Medical Center, Pediatric and Congenital Cardiovascular Surgery, Tochigi-Japan[1] Jichi Children's Medical Center, Pediatric Cardiology, Tochigi-Japan[2]

Anomalous left coronary artery (LCA) from "Right" pulmonary artery (RPA) is rare, but clinically distinct anomaly compared to usual ALCAPA syndrome with LCA of main pulmonary artery origin. Two cases, aged 5 months, 1 year and 6 months respectively, with this anomaly were encountered, the former with free-standing LCA after origin from RPA and the other with intra-mural course of proximal LCA in the posterior wall of ascending aorta before appearing in the usual retro-pulmonary space. Associated anomalies included mitral regurgitation with significant left ventricular dysfunction in the first and Chromosome 4p- syndrome with ventricular septal defect in the other. First case required LCA extension with tubed RPA segment for anastomosis to left common carotid artery and the second case repaired with unroofing technique of intra-mural portion with closure of LCA origin from RPA. Both cases showed satisfactory

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post-operative courses and post-operative studies disclosed well patent LCA in both. Mitral regurgitation disappeared and LV function normalized within 6 months without surgical intervention in the first. The second case had undergone preparatory pulmonary artery banding for associated VSD and extreme low birth weight (1506 g) without recognition of the coronary anomaly. Banding tape positioned at the bifurcation had risk of catastrophic coronary artery injury at the time of procedure or risk of ischemia after banding. LCA anatomy and configuration in these cases were NOT suitable for direct reimplantation or intrapulmonary tunneling procedure for surgical repair. This rare variant of coronary anomaly has significant diagnostic and surgical importance of recognition for successful repair.

P1901 - A RETROSPECTIVE ANALYSIS OF THE SURGICAL INDICATIONS FOR VALVE SPARING AORTIC ROOT REPLACEMENT IN PATIENTS WITH CONNECTIVE TISSUE DISORDERS ACCORDING TO THE AHA SURGICAL GUIDELINES

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Lincoln Medical and Mental Hostipal, Pediatric Cardiology-Palliative Care and Pain Management, Nyc-United States

Objective: We aim to evaluate how closely our institution follows the current recommended guidelines in regards to the timing of valve sparing aortic root replacement (VSRR).
Background: Based on the AAC/AHA guidelines for the diagnosis and management of patients with genetic syndromes with thoracic Aortic disease 2010, patients are classified in two main classes. Class I include indications for medical approach of these patient since (from the time) the diagnosis is confirmed. Class II is divided in two categories where Class IIa indicates considerations for surgical intervention in all patients with Loeys-Dietz syndrome or a confirmed TGFBR1 or TGBR2 mutation and an aortic diameter of 4.2 cm or greater by TTE or 4.4 to 4.6 cm or greater by CT and/or MRI. For women with Marfan Syndrome contemplating pregnancy, to consider prophylactic replace the aortic root and ascending aorta if diameter exceeds 4 cm. If maximal cross-sectional area in square centimeters of the ascending aorta or root divided by the patient's height in meters exceeds a ratio of 10, surgical repair is reasonable because shorter patients have dissection at a smaller size and 15% of patients with Marfan syndrome have dissection at size smaller than 5 cm.
Methods: We reviewed the charts for the last 30 patients who underwent valve sparing aortic root replacement at The Johns Hopkins Hospital and who also had a preoperative echo with aortic dimensions annotated. Patients who were followed and referred by other institution different to Johns Hopkins without a preoperative echocardiogram and patients without a conformed connective tissue disorder were excluded.
Hypothesis: Our hypothesis was that as an institution we were following the current recommendations greater than 90% of the time.
Significance: The results of this retrospective chart review will give our Pediatric Cardiology and Pediatric Cardiothoracic surgery departments insight into our current.

P1903 - SMALL DOUBLY COMMITTED VENTRICULAR SEPTAL DEFECT AND AORTIC REGURGITATION THE ERA OF EARLY SURGICAL REPAIR

Zek Sen Lim[1], Zhia Ning Lim[2], Zhen Yue Lim[3], Siew Cheng Tan[4]
UMMC, Paediatric Cardiology, Kuala Lumpur-Malaysia[1]; University of Leicester, Faculty of Medicine, Leicester-United Kingdom[2]; Cardiff Six Form College, Six Form College, Cardiff-United Kingdom[3]; UMMC, Paediatric Intensive Care, Kuala Lumpur-Malaysia[4]

Background: Aortic regurgitation is a well-known complication associated with doubly committed VSD. Lack of support at the valve and Venturi effect of the jet was believed to cause aortic valve prolapse. Early operation are normally recommended especially when the size is significant. However for small doubly committed VSDs no definitive consensus in treatment was established. Furthermore there can be difficulty in differentiating doubly committed VSDs and outlet muscular VSDs.
Methods: This is a retrospective study looking at all VSD seen in the past 3 years in University Malaya Medical Centre, a regional unit. All patients with doubly committed and outlet muscular VSDs are recruited in the study. Their images were reviewed by a pediatric cardiologist to confirm the diagnosis. Diagnosis error between doubly committed and outlet muscular were noted. The characteristics of the VSDs and evidence of prolapse were reviewed. The time interval when aortic regurgitation occurs is documented.
Results: There are a total of 318 patients with VSDs in the registry. 5 have incomplete data and has to be excluded. There were 8 patients (2.5%) with doubly committed VSDs and 10 (3.1%) with outlet muscular VSDs. There are no significant differences in the baseline characteristics for both group of patients. Kaplan Meier analysis suggest that small doubly committed VSDs is likely to progress to aortic regurgitation although this may take many years, unlike outlet muscular VSDs. There was also a significant proportion (60%) of outlet muscular VSDs misdiagnosed as doubly committed.
Conclusion: Small doubly committed VSDs should be closed surgically as there are risks of aortic valve prolapse and regurgitation. This effect is not seen with outlet muscular defect. Differentiating the two diagnosis is important.

P1923 - ANTI INFLAMMATORY AND CARDIOPROTECTIVE EFFECT OF NITRIC OXIDE DURING PEDIATRIC CARDIOPULMONARY BY PASS – CLINICAL RANDOMISED STUDY

Jacek Kolcz[1], Elzbieta Karnas[2], Wojciech Stycula[1], Magdalena Borowka[1], Zbigniew Madeja[2], Ewa Zuba-Surma[2], Janusz Skalski[1]
Jagiellonian University, Pediatric Cardiac Surgery, Kraków-Poland[1]; Jagiellonian University, Cell Biology, Kraków-Poland[2]

Objective: Cardio-plulmonary bypass (CPB) used for open heart surgery, induces systemic inflammatory response (SIRS) and contributes to ischemia-reperfusion injury of the cardioplegia-arrested heart. Multiple strategies have been elaborated to mitigate effects of CPB on perioperative stress response. We carried out a randomised clinical study to investigate an effect of nitric oxide (NO) inhalation into the oxygenator during CPB, in children.
Methods: Blood samples were taken from 24 patients (aged 8 days-13 years) during open heart surgery. CPB was managed according to previously reported, standard approach. Repeated measures were carried out before surgery, during the procedure, and 4, 8 12 and 24 hours after the operation. During the procedure nitric oxide (NO) was inhaled into the oxygenator according to the separate protocol in 12 patients. Relevant panel of cytokins and angiogenic factors was profiled by human proteome profiler Array Kits and quantitively analyzed by milliplex map kit assay. Based on screening, following analytes were selected for high-sensitivity quantative analysis: IL-10, IL-1b, TNF-a, GM-CSF, IL-6, IL-8, SDF-1, VEGF, IL-1ra, MMP-8, pentraxin-3, NT-proBNP, CK-MB, troponin I, TIMP-4, angiopoietin-2, insulin, lectin.

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Glucose and lactates were additionally measured. Repeated measures Friedman ANOVA was used for statistic evaluation of data. *Results:* Inhalation of NO decreased concentration of TnI and CK-MB and reduced level of IL-6, IL-8 and Pentraxin-3. Anti-inflammatory IL1-ra and TIMP 4 was significantly increased in NO group. SDF 1 and GM-CSF were also significantly increased in particular time points. NO-patients had lower post-bypass concentration of lactates and glucose and higher levels of Insulin, leptin and angiopoetin 1 in postoperative period. All above mentioned effects were significant at $p < 0.05$ in post hoc tests. *Conclusions:* Intraoperative NO inhalation into the oxygenator during pediatric cardiopulmonary by-pass can have anti-inflammatory, cardioprotective and reliving-metabolic-stress effects.

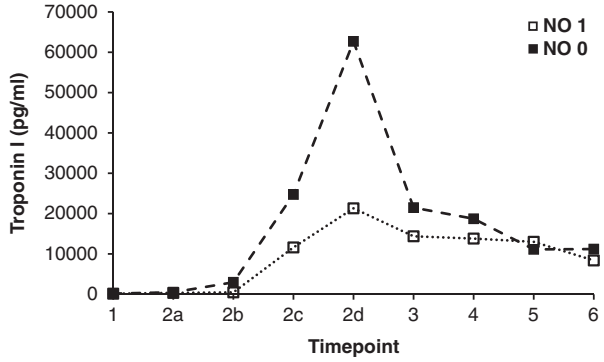


Figure 1.

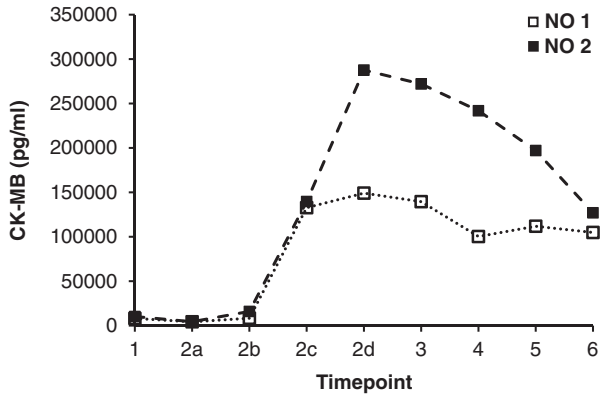
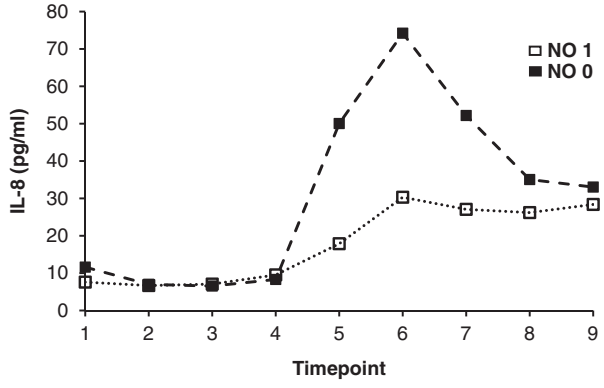


Figure 2.



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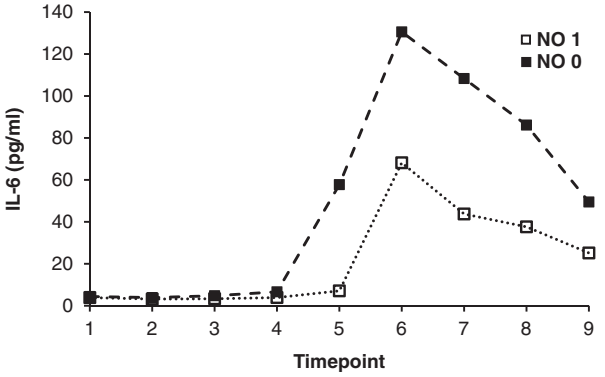

Figure 4.

P1930 - THE EFFECT OF MODIFIED BLALOCK TAUSSIG SHUNT ANASTOMOSIS ANGLE AND PULMONARY ARTERY DIAMETER ON PULMONARY FLOW

Ahmet Arnaz[1], Senol Piskin[2], Yusuf Yalçinbaş, Kerem Pekkan[2], Tayyar Sarioglu[1]

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Background: Modified Blalock-Taussig shunt (MBTS) is a palliative procedure for patients with severe cyanotic congenital heart disease associated with reduced or absent pulmonary blood flow. In this case, it was aimed to find out the best graft to pulmonary artery (Pa) anastomosis angle measuring the pulmonary blood flow, wall shear stress and shunt flow which are invariable in shunt diameter (Figure 1).
Material and Methods: Computational model of Tetralogy of Fallot with pulmonary atresia was studied using three-dimensional geometries of cardiovascular system. Three different MBTS anastomosis angle configurations connecting the right subclavian artery to the right Pa were parameterized (60 o lean to the left Pa, 90 o and 60 o lean to the right Pa) on three different Pa diameter configurations. Velocity and wall shear stress were analysed in the PTFE shunts. Flow rates at the right and the left Pa's were calculated for all configurations for comparison.
Results: In case of equal 4 mm diameter arteries, vertical shunt configuration produces the least flow rate in both pulmonary arteries. This can be explained by stagnation point at the anastomosis region of shunt and right pulmonary artery. When the left Pa diameter is doubled in size, flow going into right Pa is almost the same with same Pa diameter configuration while left Pa flow increases for all shunt angle configurations. For the third diameter configuration of 8 mm of right Pa and 4 mm of left Pa, flow going into right Pa increases slightly in case of vertical or leaned left shunt configurations (Table 1).

Table 1. Percent of total puknonary flow spirt ratios for all cases sttxied. All values are normalized by dividing by the mawmum total flow (vertical shunt, diameter of 4 mm - 8 mm).

RPA-LPA diameters	4 mm - 4 mm	4 mm - 8 mm	8 mm - 4 mm
Vertical (90*)	88	100	97
Lean right (60*)	88	88	93
Lean left (60*)	91	95	90

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Conclusions: Anastomosis angle between conduit and pulmonary arteries have crucial effect on flow splits directed to pulmonary arteries. Vertical anastomosis configurations increase the total Pa flow and thus those configurations are preferable compared to leaned anastomosis conduit configurations..

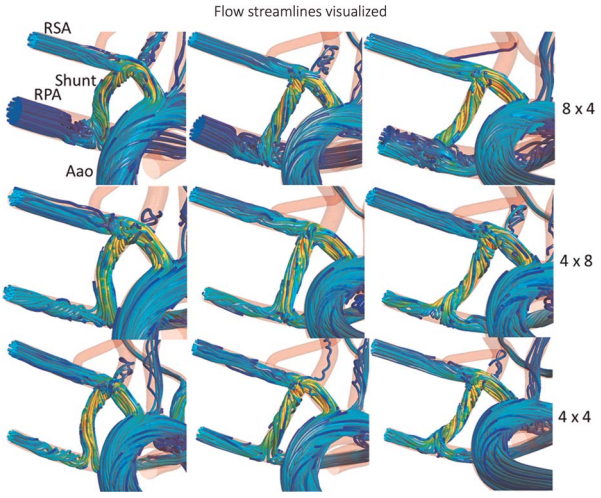

Figure 1.

P1938 - GROWTH OF LEFT VENTRICULAR OUTFLOW TRACT AND PREDICTORS OF FUTURE REINTERVENTION AFTER REPAIR OF VENTRICULAR SEPTAL DEFECT AND AORTIC ARCH OBSTRUCTION

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Kacc, Pediatric Cardiac ICU, Riyadh-Saudi Arabia[1]; Kacc, Pediatric Cardiology, Riyadh-Saudi Arabia[2]

Ventricular septal defect and aortic arch obstruction is usually associated with a narrow left ventricular outflow tract. The aim of the study was to analyze the growth and predictors of future obstruction of left ventricular outflow tract after surgical repair of these cases.
Methods: Retrospective review of patients who underwent repair of ventricular septal defect and aortic arch obstruction (coarctation or interrupted aortic arch) between July 2002 and June 2013. Echocardiographic data was reviewed and the need for reintervention was evaluated.
Results: A total of 89 patients were included. A significant left ventricular outflow tract growth was noticed after surgical repair. Preoperatively mean left ventricular outflow tract Z-score was -1.46 + /-1 (range -5.5 to 1.1) and raised to a mean Z-score of -0.7 + /-1.3 (range -2.7 to 3.2) at last follow-up ($p = 0.0001$); demonstrating relevant growth of left ventricular outflow tract after repair of ventricular septal defect and aortic arch obstruction. After primary repair 11 patients (12.3%) required reintervention with surgical repair for left ventricular outflow tract obstruction after a mean period of 36 + /-21 months. There were no significant differences in age, weight and indexed aortic valve and left ventricular outflow tract measurements between those who developed obstruction and those who did not.
Conclusion: Significant left ventricular outflow tract growth is expected after repair of ventricular septal defect and aortic arch obstruction. Small aortic valve and left ventricular outflow tract at diagnosis are not risk factors to predict the need of surgical reintervention for left ventricular outflow tract obstruction in future.

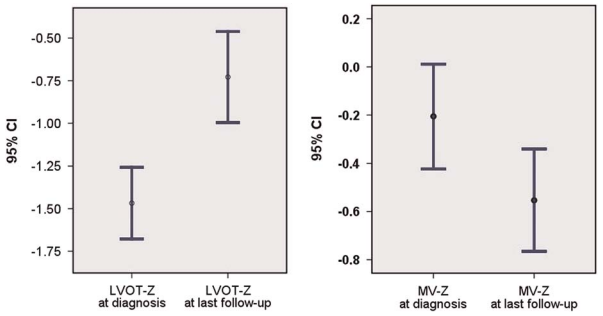

Figure.

P1946 - LONG TERM TRANSPLANT FREE SURVIVAL FOLLOWING REPAIR OF TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION (TAPVC) A STUDY FROM THE PEDIATRIC CARDIAC CARE CONSORTIUM (PCCC)

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Long-term survival, risk of transplantation, and causes of death for infants undergoing early repair of TAPVC remain ill defined. We attempted to link an international congenital cardiac surgical database of patients undergoing correction of TAPVC to two national registries, the National (USA) Death Index (NDI) and the United Network for Organ Sharing (UNOS), to understand long-term outcomes in children surviving TAPVC repair. We identified 811 survivors within the PCCC that had undergone TAPVC repair before one year of life and had sufficient personnel identifiers for linkage with the NDI and UNOS. Overall late mortality or need for transplantation was 9.3% with median follow-up of 18.6 yrs. The risk of mortality/transplant following TAPVC repair was highest in the first 18 months following hospital discharge. Comparison between patients with complex or simple TAPVC is provided in Table I. Parametric model of transplant-free survival for TAPVC conditioned to hospital discharge is presented in Figure I. Cardiac causes account for 47% of deaths for patients with simple TAPVC and 65% for those with complex TAPVC. The

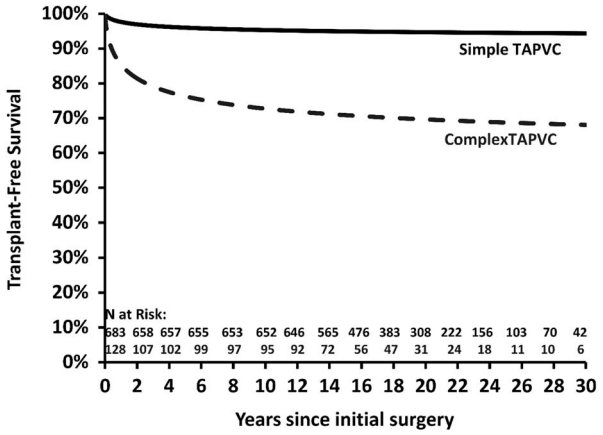

Figure 1.

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multivariable parametric regression models for transplant-free survival demonstrated that complex TAPVC (HR:5.07, 95% CI: 3.04-8.45), mixed TAPVC (HR:2.81, 95% CI: 1.56-5.03) and postoperative length of stay were associated with increased risk of death or transplant. Surprisingly, birth and surgical weight, age at repair, preoperative obstruction, and need for emergent surgery did not affect long-term transplant-free survival. Transplant-free survival after TAPVC repair is excellent with most deaths or transplant events occurring during the first postoperative year. Factors associated with worst long-term outcomes include: complex TAPVC, mixed TAPVC and prolonged postoperative LOS. Risk factors important during the immediate post-operative period had no significant effect on long-term outcomes.

Table 1. PCCC Variables, Post-Discharge Mortality and Transplantation

PCCC Variables	Overall (N = 811)	Simple[1] (N = 683)	Complex[2] (N = 128)	P-value[3] (95%)
Male	507(62 5%)	436 (63 8%)	71 (55.5%)	
Birth Weight :kg) (n = 540) median (IQR)	3 26 (2.90–3.64)	327 (2.9–3.68)	3.1 (2.84–3.53)	0.104
Surgical Weight (kg), (n = 738) median (IQR)	3.68 (3.20–4 50)	3.64 (3.20–4.40)	3.7(3.13 – 5.05)	0.193
Age at Repair (days), median (IQR)	20 (5– 80)	19 (5-72)	30 (6–165)	0.004
Anatomic Type				0.152
Supracardiac	357(44%)	302(44.2%)	55 (43%)	
Cardiac	162 (20.0%)	142 (20 8%)	20 (15.6%)	
Intracardiac	183 (22.6%)	152 (22 3%)	31 (24.2%)	
Mixed	92 (11.3%)	76 (11.1%)	16 (12.5%)	
Undefined	17 (2 1%)	11 (1.6%)	6 (4.7%)	
Emergent repair (≤2 days)	139 (17.1%)	117 (17.1%)	22 (17.2%)	0.987
Obstruction[4]	206 (25.4%)	170 (24.9%)	36 (28.1%)	0.034
Post-operative length of stay[5] (days), (n = 749) median (IQR)	11 (7 –18)	10 (7–16)	17 (9–29)	<0.001
Duration of Follow-up[6] (years), median (IQR)	-18 6 (14.5–23.0)	19.2 (15.1 –23.4)	15.3 (12.6–20 3)	<0.001
Post Discharge Mortality	66 (8.3%)	34 (5.0%)	32 (25.0%)	<0.001
Post Discharge Cardiac Tx	9 (1.1%)	1 (0.1%)	8 (6.3%)	<0.001

[1]Simple TAPVC (including PDA and ASD only),
[2]Complex TAPVC (including Associated Cardiac Anomalies)
[3]P-value refers to comparison between simple and complex TAPVC
[4]Obstruction as defined by the individual centers
[5]Post-operative length of stay was defined as date from surgery to discharge from PCCC
[6]Follow-up was defined as date of surgery until date of death or 12/31/2014.

P1953 - MID TERM OUTCOMES OF REPEAT RIGHT VENTRICULAR OUTFLOW TRACT RECONSTRUCTION USING A STENTLESS PORCINE AORTIC ROOT BIOPROSTHESIS

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Background/Hypothesis: A stentless porcine aortic root bioprosthesis has been primarily used in our institution for right ventricular outflow tract (RVOT) reconstruction in congenital heart disease. We hypothesize its longevity is similar to other conduits for this indication.
Materials and Methods: We performed a retrospective review of all RVOT reconstructions using a porcine aortic root in our institution. Ross procedure, loss of follow-up within a year of surgery, and non-cardiac related deaths were excluded. Survival and re-intervention, either by surgery, transcatheter valve implantation, balloon valvuloplasty, or bare metal stent placement were recorded. Factors associated with reintervention were assessed using Cox regression.
Results: Between January 2002 and December 2015, there were 163 patients meeting inclusion criteria. The median age was 12.2 years (range 0.7 to 36.9), median weight was 39.1 kg (range 6.9 to 176), and median body surface area 1.23 m2 (range 0.4 to 2.7). Ninety-three (57%) patients had tetralogy of Fallot. The median follow up was 5.3 years (range 1.04 to 13.7). There were no operative or cardiac related deaths. Thirty eight patients (23%) required reintervention. The rate of freedom from reintervention was 93.2% (95% CI 86.7-96.6%) at 5 years and 48.4% (95% CI 34.9-60.6%) at 10 years. Age <10 years, weight <39 kg, and body surface area <1.2 m2 at the time of valve placement, as well as valve size ≤25 mm were significantly associated with need for earlier reintervention. Sex, orthotopic versus heterotopic position, diagnosis, or concomitant pulmonary artery augmentation were not associated with earlier reintervention.
Conclusions: The porcine aortic root in the RVOT is associated with excellent survival and low mid-term need for reintervention. It longevity is comparable to published data on homografts and other bioprosthetic valves..

Table.

		5-Year Freedom from Reintervention		10-Year Freedom from Reintervention	
		Rate	95% CI	Rate	95% CI
BSA	<1.2	**95.22%**	85.82-98.44%	**24.05%**	10.06-41.29%
	≥1.2	**92.22%**	80.13-97.08%	**74.05%**	54.00-86.37%
Age	<10 years	**92.47%**	81.07-97.12%	**18.01%**	6.51%-34.12
	≥10 years	**93.57%**	83.41-97.60%	**82.59%**	66.99-91.27%
Weight	<39 kg	**93.84%**	84.32-97.66%	**25.65%**	11.76-42.11%
	≥39 kg	**92.36%**	80.49-97.13%	**79.16%**	61.18-89.48%
Valve Size	≤25 mm	**92.42%**	82.66-96.79%	**33.20%**	18.43-48.74%
	27, 29 mm	**93.88%**	81.36-98.09%	**76.31%**	54.65-88.61%

P1956 - IS BICUSPID PULMONARY VALVE USING POLYTETRAFLUOROETHYLENE MEMBRANE SUPERIOR THAN BIOPROSTHETIC VALVE NOT SUPERIOR BUT NOT INFERIOR COMPARISON STUDY OF DURABILITY AND FUNCTION

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Background: The hand-sewn polytetrafluoroethylene (PTFE) bicuspid valve is good alternatives in pulmonary position. The aim

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of this study is to evaluate midterm outcome of PTFE bicuspid valve with comparison with bioprosthetic valve.
Methods: We retrospectively reviewed 149 patients who underwent pulmonary valve replacement (PVR) for chronic pulmonary regurgitation from Jan 1999 to Dec 2015. Patients were divided two groups; Hancock II porcine valve group (group 1, N = 80) and PTFE valve group (group 2, N = 69). We evaluated durability and function of valves.
Results: The mean ages at PVR were 19.4 ± 10.5 years old in group 1 and 19.6 ± 7.8 years old in group 2 (p = 0.909). The median valve sizes were 25mm (range; 19 ~ 27mm) in group 1 and 28mm in group 2 (range; 20 ~ 30mm) (p = 0.000). There were 1 early hospital death in group 2 due to cardiac dysfunction and 1 late death in group 1 due to heart failure. The median follow up durations were 8.6 years (range; 0 ~ 15 years) in group 1 and 2.9 years (range; 0 ~ 6.3 years). The 15 years survival rate in group 1 was 98.8% and 6 years survival rate in group 2 was 98.6% (p = 0.916). The event free rate for reoperation or catheter intervention were 83.2% at 10 years and 72.1% at 15 years in group 1, and 82.8% at 6 years in group 2 (p = 0.07). For MRI analysis, the right ventricular volumes of both groups significantly decreased. However, right ventricular ejection fractions (RVEF) of both group were not improved.
Conclusion: The overall durability and survival rate of PTFE membrane in pulmonary position were satisfactory and there was no significant difference in comparison with Hancock II porcine valve. And, there was no significant difference in the effect of volume reduction and RV function.

P1957 - A CASE OF COMBINATION THERAPY WITH RENAL REPLACEMENT THERAPY AND SURGICAL REPAIR FOR IDIOPATHIC MITRAL VALVE CHORDAE RUPTURE IN AN INFANT

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Background: Idiopathic mitral valve chordae rupture is a sudden onset and life threatening disease due to severe cardiac failure. The disease was thought to be rare in young children, but several cases in infants without congenital malformations of mitral valve have been reported in Japan. Recently, the disease has gained significant attention as a cause of sudden death in infants. We encountered a case with severe cardiac failure, acute kidney injury and hepatic congestion due to idiopathic mitral valve chordae rupture, and a combination therapy with surgical repair and renal replacement therapy was effective.
Cases: A 7-month-old boy had fever and diarrhea that lasted for five days. Ten days later, he presented to a family doctor with sudden onset of paleness and tachypnea. Heart murmur and tracheal stenosis sound were heard, and the patient developed metabolic acidosis and cardiogenic shock. Severe mitral regurgitation and anterior leaflet prolapse was detected with echocardiography, and he was diagnosed with idiopathic mitral valve chordae rupture. He was intubated and air-transferred to our hospital for operative management. On arrival at our hospital, metabolic acidosis and cardiogenic shock had improved. However, the patient was in a state of acute kidney injury and severe hepatic congestion. After renal replacement therapy, the patient was operated next day. Operative findings confirmed ruptured chordae of the anterior mitral leaflet and posterior commissure. There were no structural abnormalities or signs of vegetation. Surgical procedure included mitral valve repair with artificial chordae and commissural plasty. Postoperative course was good with no recurrence of regurgitation.
Conclusion: Combination therapy with surgical repair and renal replacement therapy led to a good outcome in a patient with idiopathic mitral valve chordae rupture. Early and adequate treatment prior surgery coupled with decision on appropriate timing for surgery might be a crucial factor for the better outcome.

P1961 - CONE RECONSTRUCTION FOR EBSTEIN'S ANOMALY WHAT DEFINES THE NEED FOR ADDITIONAL BIDIRECTIONAL CAVOPULMONARY ANASTOMOSIS

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Objectives: We aimed to investigate whether the tricuspid valve (TV) diameter after "cone reconstruction" (CR) as primary biventricular, "anatomical" correction of Ebstein's anomaly decisively influences the need for additional bidirectional cavopulmonary anastomosis (BCPA, "one-and-a-half" repair).
Methods: Retrospective echocardiographic analysis of the postsurgical TV annulus was performed in patients who underwent CR between 06/2013 and 1/2017. The Z scores were calculated and correlation with the bi or "one-and-a-half" result was checked. Further, the influence of the Carpentier classification for the outcome was analysed.
Results: Thirty three CR were performed without mortality in 31 patients (median age 4 years, weight 18.6 kg), in 15 (48%) of them with and in 16 (52%) without BCPA. The postsurgical course was uneventful (mechanical ventilation in median 19 h, hospital stay 8 d) except for temporary ECMO support in one patient. Small atrial septal defect was left in 16 patients. Central venous pressure at the end of the operation was in median 12 mmHg and showed no correlation with BCPA necessity. According to the Carpentier classification (class B: 12 (23%), C: 19 (53%)) we found no correlation with regard to BCPA. Postsurgical TV regurgitation was absent in 13 (43%), low in 13 (43%) and mild in 4 (14%) patients. The mean diameter of the TV annulus and equivalent Z score were significantly lower (p = 0.002 and p = 0.01, respectively) in patients who needed an additional BCPA.
Conclusion: The resultant diameter of the TV created using CR seems to be co-decisive: in the case of Z score > - 2 the additional BCPA may be avoided even with severe dysplasia of the TV; however, it seems to be necessary with smaller TV diameter/Z score. Intraoperative TV-Z score determination after CR may be helpful in the decision-making with regard to "biventricular" or "one-and-a-half" correction of the Ebstein anomaly.

P1963 - LONG TERM FOLLOW UP AFTER ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY REPAIR

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Background: Anomalous origin of the Left coronary artery from the pulmonary artery (ALCAPA syndrome) is an uncommon congenital malformation that affects 0.008% of population.
Materials and Methods: Retrospective review of all children diagnosed with ALCAPA at our institution over the last 15 years. Surgical techniques, as well as clinical, ecocardiographic and electrocardiographic variables associated with outcomes were evaluated.
Results: 15 consecutive children were included. Initial presentation was: 8 (53.3%) incidental finding, 6 (40%) heart failure and 10 (6.6%) cardiogenic shock. Fourteen (93.3%) children had ECG abnormalities. Different surgical techniques were used: coronary reimplantation in numero (60%), Takeuchi technique in 10 (6.6%) and ligation in numero (26.6%). Mitral repair was necessary in 2 (13.3%). One patient died before surgery and there was one case of early postoperative death. Average follow-up was 5.50 ± 2.71 years. At follow-up, number (86,66%) were asymptomatic. Two major complications were identified: prosthetic mitral valve dysfunction and aortopulmonary fistula (patient treated with Takeuchi tecnique). All ecocardiographic values improved, with significant differences in EF ($p = 0.02$). Mitral regurgitation improved in most of the cases (67% of patients with moderate to severe MR at diagnosis vs 20% after one year, $p = 0{,}016$). In n (60%) of cases, normalization of ECG changes was observed.
Conclusions: Translocation of the left coronay to the aorta has proved to have good overall results, with significant clinical improvement and a progressive normalization of the LV systolic function. Correction of ECG changes and a decrease in mitral regurgitation was also observed in most of the patients.

Table 1. Ecocardiographic variables at diagnosis and one year after surgical treatment.

	At diagnosis (mean ± SD)	After one year (mean ± SD)	P
LVEDd (mm)	41.3 ± 7.3	39 ± 10,23	0.10
LVEDd (Z)	3.3 ±2.3	2.4 ± 2.1	0.08
EF (%)	46.8 ± 15.5	60.1 ± 9.0	0.02*
FS(%)	26.8 ± 8.0	33.5 ± 0.8	0.18
Degree of mitral regurgitation	n(%)	n(%)	
- Mild (%)	2 (13.3)	6 (40)	0.12
- Moderate (%)	7 (46.6)	1 (6.6)	0.03
- Severe (%)	3 (20)	2 (13.3)	1.00

LVEDd: left ventricular end diastolic. EF: Ejection fraction. FS: Fractional Shortening.

P1969 - GROSS AND HISTOLOGICAL CHARACTERISTICS OF VENOUS HOMOGRAFTS USED IN HEART SURGERY

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Background: Saphenous/femoral vein homografts are used increasingly to establish right ventricle - pulmonary artery continuity in complex heart defects.
Methods and Materials: We examined 15 of 16 vein homografts (exclusion for internal stent placement) explanted from October 2015 to December 2016 and describe their gross and histological characteristics. Explanted grafts were examined fresh and sectioned for histological examination. Slides were scored semi-quantitatively for inflammation, remodeling, neointima formation, and calcification.
Results: The vein grafts (3 saphenous, 12 femoral) were implanted in infants 1-118 days old (median 4.5 days) as part of a composite Sano shunt (11 patients) or to establish right ventricle-pulmonary artery continuity in 2-ventricle hearts (4 patients). The duration in situ was 96-389 days (median 130 days). The vein homografts were explanted for conversion to bidirectional Glenn (11 patients), distal homograft stenosis (1 patient), pseudoaneurysm at anastomosis (1 patient), or elective upsizing at other surgical procedure (2 patients). Vascularized fibrous or granulation tissue covered the grafts externally. The vein wall appeared thickened in all cases due partly to a fibrous neo-intima. The venous valve was intact in 12 cases. Histological exam showed consistent invasion of host cells (predominantly fibroblasts) into the vein wall with few inflammatory cells (macrophage, lymphocytes). Graft collagen appeared to be undergoing remodeling with deposition of new host collagen. Host blood vessels invaded the adventitia of most grafts and a mild foreign body reaction (giant cells, histiocytes) was noted in those with an external, confining stent. Calcification, mostly at suture lines, was absent or mild in 14 cases and moderate in 1 case. Neointima formation was mild in 10 and moderate in 5 cases.
Conclusion: Vein homografts appear to remodel, provoke minimal inflammation and calcification. The valve functions at least 1 year.

P1970 - GROSS AND HISTOLOGICAL FINDINGS IN BIOPROSTHETIC MITRAL VALVES EXPLANTED FROM CHILDREN UNDER 5 YEARS OF AGE

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Background: Mitral valve replacement (MVR) in very young children is challenging. We investigated the mechanisms for early bioprosthetic mitral valve failure in young patients through review of the macroscopic and microscopic findings in explanted bioprosthetic valves.
Methods: Patients who underwent MVR with a bioprosthetic valve at Boston Children's Hospital between January 2010 to April 2016 at <5 years of age were identified by searching the departmental computerized database. Valve failure was defined as prosthetic mitral valve explantation with mitral valve re-replacement. Valves were examined fresh and sections of leaflets prepared for histology.
Results: Porcine (25) or bovine pericardial (6) bioprosthetic valves were used in 31 of 77 MVRs during the study period. Valve failure (predominantly stenosis) occurred in 10 patients (32%) and was due to pannus deposition (5 porcine, 2 pericardial) and intrinsic leaflet calcification (3 porcine). Pannus formation occurred predominantly on the ventricular surface of leaflets adjacent to the posterior wall but not the leaflet near the outflow tract. In 6 of these valves thrombus was identified at the leading edge of pannus formation. Microthrombi were identified on the leaflets of another normally functioning valve studied at autopsy after 1 month in situ but not on one studied after 1 week in situ. Pannus involved the atrial surface in 2 other valves (1 porcine, 1 bovine

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pericardial) and appeared to be an extension of the fibrous tissue covering the sewing ring.
Conclusions: In addition to intrinsic calcification, excessive pannus deposition is a prominent cause of early bioprosthetic valve failure in very young patients. Pannus formation appears due to organization of thrombus deposited on leaflets and to ingrowth from host tissue contiguous to the sewing ring as part of a foreign body reaction.

P1991 - VARIATION IN PERIOPERATIVE CARE ACROSS CENTERS FOR INFANTS UNDERGOING THE STAGE II PALLIATION FOR HYPOPLASTIC LEFT HEART SYNDROME

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Background: In the Single Ventricle Reconstruction (SVR) Trial, neonates with hypoplastic left heart syndrome were randomized to a modified Blalock-Taussig or right ventricle-pulmonary artery shunt. They otherwise received local standard of care. We analyzed practice variation at Stage II (S2) surgery.
Methods: Perioperative data were collected prospectively and available in the SVR public use database. Practice variations across the 14 centers are described for 397 subjects who underwent S2. All data are center-level specific and reported as median, interquartile (IQR) and full range unless otherwise specified.
Results: The median age and weight at S2 was 5.4 months (IQR 4.9-5.7; full range 4.0-6.6) and 5.7 kg (5.5-6.1; 5.1-6.7) with 30% performed urgently/emergently. Prior to S2, a majority had a catheterization (IQR 98.5-100%), digoxin was used by 11 centers in 25% of subjects (23-31%; 13-50%), and 81% were on oral feeds (68-84%; 63-95%). In 12/14 centers, 85% of subjects (58-90%; 22-100%) had a bidirectional Glenn and 5 centers used a hemi-Fontan in 55% (33-92%; 18-100%). Cardiopulmonary bypass time was 96 minutes (75-113; 40-130). In aggregate, 26% (103/397) of subjects had deep hypothermic circulatory arrest (DHCA) >10 minutes. In 13/14 centers using DHCA, a median of 12.5% of subjects had DHCA >10 minutes (8-32%; 4-100%). In 7/14 centers, 5% of subjects (2-40%; 1-70%) were extubated in the operating room. Postoperatively, ICU length of stay (LOS) was 4.8 days (4.0-5.3; 3-10) and total LOS was 7.5 days (6-10; 5-14). At 11/14 centers, 19% of subjects (12-25%; 2-83%) were discharged on oxygen with a median saturation of 83% (80-84%; 79-85%).
Conclusions: In the SVR trial, practice varied widely among centers for nearly all perioperative factors surrounding S2. These findings are consistent with variation previously reported for the Norwood procedure. Further analysis may facilitate establishing best practices by identifying the impact of practice variation.

P1994 - LONG TERM OUTCOMES OF FONTAN PROCEDURE IN PATIENTS WITH ATRIOVENTRICULAR VALVE DISFUNCTION

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Background: Atrioventricular valve (AVV) regurgitation management still a challenge in patients with single ventricle physiology and is one of the predictors of adverse outcomes after the Fontan procedure (FP).
Methods: We studied patients who underwent FP and concomitant valve surgery between 1984 and 2015. Factors associated with patient outcome were investigated retrospectively.
Results: A total of 420 patients underwent FP. Sixty-four (15.2%) patients had AAV regurgitation, and 36 (8,6%) underwent concomitant valve surgery (valve repair = 31; valve replacement = 5). Previous AAV regurgitation (hazard ratio [HR], 3.499; 95% confidence interval [CI], 1.566 to 7.817; $p = 0.002$), concomitant valve repair (HR, 3.334; 95% CI, 1.352 to 8.224; $p = 0.009$) and valve replacement (HR, 9.358; 95% CI, 2.205 to 39.720; $p = 0.002$) were risk factors for mortality.
Conclusion: In our series, a greater number of patients with AAV regurgitation were treated simultaneously with the FP. Performing concomitant valve repair/replacement was a risk factor, so whenever possible these procedures should be done during the bidirectional Glenn shunt to improve the selection of the candidates to complete the FT operation.

P2000 - REPAIR OF LATE PRESENTING INTERRUPTED AORTIC ARCH WITH SEVERE PULMONARY HYPERTENSION USING A PULMONARY AUTOGRAFT TUBE

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Background: The optimal surgical technique for repair of interrupted aortic arch (IAA) remains controversial. We present a case of late-presenting IAA with severe pulmonary hypertension which was repaired using a pulmonary autograft tube.
Materials and Methods: A 34-month-old boy from a foreign country was admitted to our hospital for management of his congenital heart disease. Echocardiography revealed type A IAA with a large perimembranous ventricular septal defect (VSD) and severely dilated main pulmonary artery. Cardiac catheterization was performed and pulmonary vascular resistance was calculated as 8.9 units at room air. It fell to 5.8 units with 100% inspired oxygen. The VSD was closed using a bovine pericardial patch with a 5.6 mm fenestration. An autograft tube of diameter 12 mm was made of anterior wall of the main pulmonary artery. The arch was repaired by interposing the pulmonary autograft tube between the descending thoracic aorta and the ascending aorta under selective cerebral perfusion.
Results: The patient could not be weaned from cardiopulmonary bypass because of pulmonary hypertension and right ventricular dysfunction. He underwent extracorporeal membrane oxygenation support for 3 days. Postoperative course was complicated by

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mediastinal bleeding and small amount of subdural hematoma. There was no neurologic deficit persisting at discharge. Echocardiography at discharge revealed left-to-right shunt through a VSD fenestration and normal biventricular systolic function. Postoperative computed tomography showed wide patent aortic arch with natural geometry (Figure).

Conclusions: Repair of IAA using a pulmonary autograft tube can be a good option in selected patients because of its ability of maintaining natural arch geometry, growth potential, and possible beneficial effect in preventing left main bronchus compression.

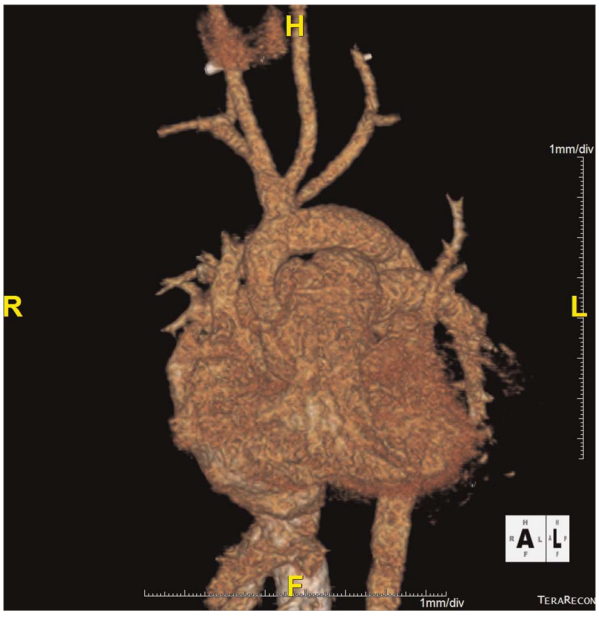

Figure.

P2004 - UNUSUAL CONGENITAL GERBODE DEFECT ASSOCIATED WITH DOWN SYNDROME; RARE CASE WITH SUCCESSFUL COMPLEX HEART SURGERY REPAIR IN THIRD WORLD HOSPITAL

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Gerbode defects are left ventricular (LV) to right atrial (RA) communications, a rare case (0.08%) which could be congenital or acquired. Can result from either direct communication between LV and RA, or indirect connection from a membranous ventricular septal defect (VSD) and a defect in the tricuspid valve leaflet. 76% of all congenital heart disease in neonates correlates with Down syndrom. We hereby, report one rare case of indirect type III Gerbode defect from a 1 years old baby boy with down syndrome presented with dyspnoe, difficulty of breast feeding and failure to thrive. History of ANC regularly in obstetrician, already known for heart abnormalities during 7 months of pregnancy. Normal baby labour, aterm with birth weight 3300 grams. Clinical examination revealed a pansystolic murmur and grade 2 parasternal heave. Echocardiogram showed 3,4 mm and 7,8 mm of perimembranous VSD with left-to-right shunt (2 jet) and small ASD with left-to-right shunt measuring 5 mm in diameter. Came to our hospital for surgery repair after been diagnosed in abroad. Under cardiopulmonary bypass, bicaval cannulation and right atriotomy, showed there were a perimembranous VSD measuring 10 mm in diameter, defect from LV to RA measuring 5 mm in diameter known as Gerbode defect lies beneath the septal leaflet of tricuspid valve. Underwent successful closure defect with 0,4 mm goretex pacth of his VSD and Gerbode defect using 5–0 prolene with pledgets and followed by primary ASD closure by leaving residual ASD 2 mm. Septal tricuspid leaflet was free with no perforation. Tricuspid valve was tested using saline injection into the RV and found to be competent. Hospitalized for one week and became healthy. He was doing well at 18-months of follow up.

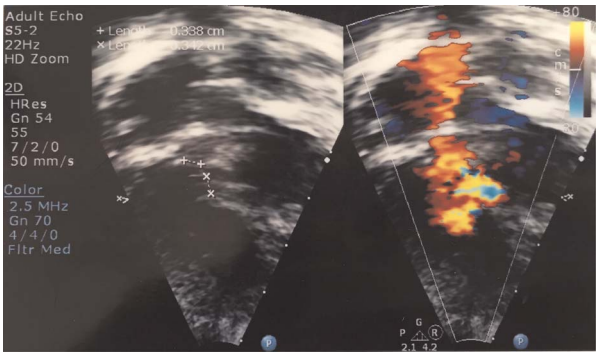

Figure.

P2006 - SHORT TERM OUTCOME OF THE OF ASD CLOSURE BY RIGHT VERTICAL AXILLARY THORACOTOMY A SINGLE CENTRE EXPERIENCE

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Background: This prospective observational study aims to evaluate the outcome of Atrial Septal Defect closure by right vertical axillary thoracotomy in terms of safety, reliability, reproducibility and cosmetic result.

Methods: The study was conducted in the department of Cardiac Surgery at Ibrahim Cardiac Hospital and Research Institute Dhaka during the period of May 2016 to January 2017. Within this 9 months period total 20 cases of ASD closure done by right vertical axillary thoracotomy. 12 were female and 8 were male. Age ranged from 4 yrs to 30 yrs (Mean 12.35 yrs). Body weight ranged from 12 kg to 58 kg. (Mean 25 kg). Outcome was assessed in terms of Total duration of CPB, Aortic occlusion time, Total operating time, volume of drainage, ICU stay, Duration of ventilation, Total hospital stay, Length of incision.

Result: Mean duration of CPB and aortic occlusion time was 50 min and 35 min respectively. Mean total operating time was 200 min. Mean duration of ventilation, ICU stay and hospital stay were 3 hrs and 18 hours and 7 days respectively. Drainage were minimal and Chest drains removed on 1st POD in all the cases. No blood transfusion required in any case. Mean length of incision was 5.5 cm. No mortality or morbidity or wound related complications observed. No narcotic analgesic required after operating day. Psychological and cosmetic results were excellent. There was no residual shunt in the follow up. echocardiogram.

Conclusion: ASD closure by right vertical axillary thoracotomy is equally safe and reproducible maneuver with superior cosmetic and psychological outcome than all other surgical technique and can be practiced in selected cases where device closure is not suitable and in patients who are not agreed to accept Device closure.

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P2008 - REPAIR OF TETRALOGY OF FALLOT WITH VALVE AND ANNULUS PRESERVATION BASED ON MORPHOLOGY OF LEAFLETS – EARLY FOLLOW UP

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Background: Conventional repair of TOF is based on size of the pulmonary annulus. We addressed the repair purely based on the morphology of the leaflets of pulmonary valve. If the leaflets are supple and pliable, we preserved the annulus aggressively irrespective of the Z-score to avoid pulmonary regurgitation.
Methods: From March 2013 to December 2016, 138 patients underwent repair of TOF with annulus preservation using the valve sparing technique. The age ranged from 4 months to 6 years (median 18 months) and the weight from 4.2 to 17 kg (median 8.4 kg). 60 patients had a pre operative Z score $\leq$ -3. The infundibular resection was done from the trans-atrial side and then the pulmonary artery was opened to inspect the valve. If the valve was salvageable, the commissural fusion was carefully split with sharp blade and the subvalvular tissue was aggressively resected with sharp scissors all around the RVOT. The main pulmonary artery was augmented with a pericardial patch.
Results: There were one death due to complications associated with infection. The median follow up was 20 months. Three patients who had a pre operative Z score of < -3 developed significant obstruction requiring re-operation. Two patients required a trans-annular incision and RVOT reconstruction with PTFE valve. One patient required further resection of the infundibular muscle. Twelve patients developed moderate gradient (30–40 mmHg) at the infundibular level and are on close follow up. The remaining 122 patients have gradients $\leq$ 25 mmHg at the infundibular level.
Conclusions: Preservation of the annulus during repair of TOF prevents pulmonary regurgitation and protects the right ventricular function. Morphology of the pulmonary valve leaflets determines the decision to preserve the annulus and valve function in TOF repair irrespective of the Z-score.

P2012 - OPTIMAL PEDIATRIC MYOCARDIAL PROTECTION SAME GOAL DIFFERENT WAYS

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Background: Myocardial protection in pediatric population during heart surgery is still a matter of considerable debate. Looking for the best cardioplegic strategy, we surveyed congenital heart surgeons about their practice.
Methods: A questionnaire was e-mailed to 88 institutions and we recieved 33 responses, which were analyzed. The survey adressed the type of cardioplegia, dosage, administration, pressure and difference in cyanotic/acyanotic patients. The respondents had to choose among several predefined response options. All percentages are based on the number of the entire reporting cohort (n = 33).
Results: Almost half of surgeons (45%) use more than one type of cardioplegia. A wide variety of cardioplegic solutions were described. More than a half (52%) use both crystalloid or blood-based cardioplegia in different situations. Induction and maintance dosage depend on the type of cardioplegia. There is also difference in calculation of the dosage with 12% of the surgeons using BSA, and another 12% measuring exposure time. When using more than one type, CUSTODIOL (®) is generally associated with long and complex operations. The majority (n = 18) of surgeons prefer delivery pressure between 30-50 mmHg, measured in the aortic root. None of the respondents use different cardioplegia in cyanotic/acyanotic patients.
Conclusions: This survey demonstrates that myocardial protection strategies differ greatly in type of solution, pressure and even in measuring methods from one surgeon to another. More studies associated with postoperative markers of the different strategies should be considered.

P2014 - RIGHT VENTRICULAR OUTFLOW RECONSTRUCTION WITH HANDMADE VALVE CONDUIT – A SHORT EXPERIENCE

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Background: Right ventricular outflow tract continuity abnormalities are one of the most commonly encountered entities in the field of congenital cardiac surgery. Various strategies including homograft, valve conduit, Contegra, and patch enlargement with valve replacement or reconstruction are used to restore anatomical and functional continuity between right ventricle and pulmonary artery. In countries like Pakistan these may not be easily available and affordable. We report the experience of our short observational study of using a handmade trileaflet valve conduit to reconstruct the right ventricular outflow tract.
Materials and Methods: From September 2015 to December 2016, a total of 15 patients with different congenital heart disease diagnoses in the pediatric age group (up to 16 years) at the Aga Khan University Hospital underwent open-heart surgery including restoration of RV to PA continuity by using handmade valve conduit. The size of the conduit was determined by using an available nomogram. The bovine pericardium and thin sheet PTFE sheets (0.5mm) were used to construct conduit and valve respectively.
Results: Patients ranged from 1 year to 16 years. Seven patients have had previous palliation including 4 BT Shunts and 3 PA banding. One patient underwent 3rd time redo procedure for RV to PA homograft stenosis. Postoperative complications were observed in 4 patient including 2 in hospital deaths and 2 required interventions. One patient developed aneurysm at RV- conduit junction requiring surgical repair and the other underwent conduit dilatation for moderate to severe stenosis (gradient 60mmg). No significant regurgitation was observed in this series. Overall postoperative gradients were stable with mean gradient 25.3 mmhg (8mmhg – 60mmhg).
Conclusions: This short report highlights that the handmade valve conduits are a cost effective alternative in this part of the world where well-established conduits have cost implications and questionable availability.

P2017 - STAGED HYBRID CLOSURE OF PATENT DUCTUS ARTERIOSUS IN PATIENTS WITH PATENT DUCTUS ARTERIOSUS AND SEVERE PULMONARY ARTERIAL HYPERTENSION

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Background: The purpose of the study is to evaluate outcomes of staged hybrid closure of patent ductus arteriosus (PDA) in patients with PDA and severe pulmonary arterial hypertension (PAH).

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Materials and Methods: From January 2007 to December 2016, 12 patients with PDA with severe PAH underwent surgical partial closure of PDA. Medical records of them were reviewed retrospectively.
Results: The mean age of patients was 11.0 ± 10.2 years old (range: 0.9 ~ 36.6, median: 7.5 years old). The mean preoperative right ventricular (RV) to left ventricular (LV) pressure ratio was 0.87 ± 0.11 (range: 0.67 ~ 0.97, median: 0.93). The mean preoperative pulmonary vascular resistance index was 8.9 ± 3.7 Wood units (WU) (range: 3.8 ~ 14.4, median: 8.6 WU). During the follow-up, there was no mortality and no symptoms associated with right heart failure. Eight patients underwent staged complete closure of PDA percutaneously after a mean follow-up of 17.8 ± 8.4 months (range: 10.1 ~ 33.7, median: 14.1 months). The RV to LV pressure ratio was significantly reduced to a mean 0.56 ± 0.08 (range: 0.44 ~ 0.67, median: 0.52) and pulmonary vascular resistance index was also significantly reduced to a mean 6.0 ± 2.0 WU (range: 2.5 ~ 8.3, median: 6.2 WU). Two patients wait for complete closure and 2 patients were lost to follow-up.
Conclusions: Staged hybrid closure of PDA is a safe treatment option in patients with PDA and severe PAH.

P2034 - MODIFIED BLALOCK TAUSSIG SHUNT COMPARISON BETWEEN STERNOTOMY AND THORACTOMY IN A SINGLE CENTER

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Background/Hypothesis: To compare the results in terms of morbidity and mortality between patients who received a modified Blalock-Taussig Shunt (BTS) performed by thoracotomy or by sternotomy under cardiopulmonary bypass (CPB).
Material and Methods: Retrospective review. We analyzed all the patients younger than 3 months who needed a BTS to assure pulmonary flow between 2005 and 2016. We excluded patients following a Norwood operation. In this period, 33 BTS were performed in our centre: 13 (39,4%) were performed by sternotomy under CPB (Group A) and 20 (59,6%), by thoracotmy (Group B).
Results: Pre-operative characteristics are shown in Table 1. In-hospital mortality was 6% (7,1% in group A and 5,3% in group B). 6 patients (18%) developed acute thrombosis of the BTS (7,8% in group A and 26% in group B). Average time of ICU, mechanical ventilation and hospitalization are shown in Table 2. 70% of patients required inotropes (85% in group A and 60% in group B). No significant differences were observed between the two groups regarding the postoperative outcomes. Median follow-up was 38 months (Range 7-80 months). Survival after follow-up was 84% (91,7% in group A and 78,9% in group B). Next-step surgery was achieved in 57,6% of patients (38,5% in group A and 75% in group B). 27,3% of patients needed stenting of pulmonary artery (7,7% in group A and 40% in group B, p value <0,05) 15,1% of patients needed ductus stenting due to BTS failure (0% in group A and 25% in group B, p value = 0,01).
Conclusion: We consider that midline approach under CPB is safer than thoracotomy, due to a lower risk of acute thrombosis and inferior rate of ductus stenting or pulmonary artery intervention during the follow-up..

Table 1.

Table 1	Global	Group A	Group B
Age (days)	42 ± 31	38 ± 30	45 ± 32
Weight (kg)	3,3 ± 0,6	3,2 ± 0,7	3.4-0,6
Age under 30 days	44%	50%	40%
Single Ventricle	34%	35%	33%
BTS/Weight index	1,16 ± 0,15	1,15 ± 0,15	1,18 ± 0,15
Aristotle Score (comprehensive)	8 ± 2,8	9,0 ± 3,7	7,5 ± 1,9

Table 2.

	Global	Group A	Group B
Mortality	6%	7,1%	5,3%
BTS thrombos is	18	7,8%	26%
ICU (days)	15 ± 16	17 ± 17	13 ± 15
MV (hours)	108 ± 29	125 ± 179	98 ± 142
Hospitalization (days)	25 ± 22	32 ± 25,5	20 ± 19,5

P2040 - AN APPRAISAL OF MATERIALS AND VALUE FOR PATCH RECONSTRUCTION OF THE PULMONARY ARTERIES

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Background/Hypothesis: We sought to evaluate quality (re-intervention) and cost of various patch materials used for main and branch pulmonary artery (PA) patch reconstruction.
Patients/Methods: We conducted a retrospective chart review of all patients undergoing patch reconstruction of the main or branch PAs (w/wo other cardiac surgery) at our institution over the interval of 1990-2015. We analyzed 5 patch materials: CM – porcine submucosal matrix; BovP – bovine pericardium; BPH – branch patch pulmonary homograft; G – goretex; and AutP – autologous pericardium. Primary outcome was re-intervention on the reconstructed PA site (not number of patients). Median follow-up was 5.3 years (IQR 2.5-9.5).
Results: The cohort included 341 patients and 442 patch reconstruction sites. Median age (IQR) and weight were 0.53 yrs (0.3-1.4) and 6.9 kg (5.1-8.9). TOF/PS and other biventricular repair constituted 54.3% of the cohort. Patch sites included main PA (w/wo) extension to branch for 143 patches (32.4%); L PA and hilar for 90 (20.4%); R PA and hilar for 105 (23.8%); and both branches for 65 (14.7%). Discharge and late mortality occurred in 4 (0.8%) and 12 (2.6%) of patients. Surgical or catheter-based re-intervention occurred as primary indication for 66 patches (14.9%); another 68 patches received re-intervention which may not have occurred had another major procedure not been indicated. Intervention and cost per material data are shown in Table 1 – notably for AutP re-intervention occurred in 11.5% of patches versus 30.6% of BPH patches, p = 0.0003.

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Conclusions: For PA reconstruction, there is considerable variability in types of patches, unit cost per patch, and material-related re-intervention. Autologous pericardium is free and provided the lowest rate of patch re-intervention.

Table 1. Re-intervention and Costs for PA Reconstruction Patches

Type of patch used at index procedure	Patch Unit Cost	Total number of patches used	Number of patches needing re-intervention	Re-intervention (%)	Total Cost
Porcine submucosal matrix (CM)	$795	65	11	16.9	$51,675
Bovine pericardium (BovP)	$500	63	23	36.5	$31,500
Branch patch pulmonary homograft (BPH)	$6397	212	84	39.6	$1,356,164
Goretex (G)	$581	34	10	29.4	$19,754
Autologous pericardium (AutP)	$0	61	7	11.5	$0

P2051 - INTERMEDIATE TERM FOLLOW UP AFTER TRANSCATHETER FENESTRATION CLOSURE IN PATIENTS WITH FENESTRATED FONTAN PROCEDURE; SINGLE CENTER EXPERIENCE

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Samsung Medical Center, Sungkyunkwan University School of Medicine, Pediatrics, Seoul-Korea, South[3] Samsung Medical Center, Sungkyunkwan University School of Medicine, Thoracic And Cardiovascular Surgery, Seoul-Korea, South[4]

Objective: To review and compare the clinical and catheterization data before and after device closure of fenestration on patients treated with fenestrated Fontan procedure.
Methods: The clinical records were reviewed for demographic, laboratory and hemodynamic data of patients who underwent follow up cardiac catheterization after fenestration closure.
Results: Thirty two patients underwent fenestration closure from 1998 to 2009 at Samsung medical center. Twenty four asymptomatic patients underwent follow up cardiac catheterization at median 8.8 years after closure (range: 4.4-12.5). Median age at follow up catheterization was 13.2 years (range: 10.7-21.8). In theses 24 patients, various devices including detachable coils($n=8$), Amplatzer duct occluder ($n=2$) and Amplatzer vascular plug I ($n=14$) were used for fenestration closure at median interval of 19 months after Fontan operation (range: 8-107). Twenty two patients had aspirin medication. Systemic ventricular morphology was left ventricle in 11 patients, right ventricle in 10 patients and undetermined morphology was in 3 patients. On follow up angiography, fenestration leakage was noticed in 4 patients who underwent fenestration closure with coil. Mean pulmonary artery pressure(PAP) was 10.9 ± 2.3 mmHg at fenestration closure and 13.7 ± 4.5 mmHg at follow up ($p=0.003$). Pulmonary vascular resistance was 1.97 ± 0.93 wood units at closure and 1.63 ± 0.7 wood units at follow up (p-value $=0.08$). Mean ventricular end-diastolic pressure(VEDP) was 7.0 ± 3.2 mmHg at closure and 11.3 ± 3.4 mmHg at follow up ($p<0.01$) Mean VEDP at follow up was correlated with duration of Fontan circulation ($r=0.35$, $p=0.023$), interval between closure and follow up ($r=0.333$, $p=0.029$), and mean PAP at follow up ($r=0.477$, $p=0.006$).
Conclusions: There was statistically significant change in mean PAP and mean VEDP at median 8.8 years follow up after fenestration closure. It is unclear whether these changes are due to natural course of Fontan surgery or is due to the result of fenestration closure.

P2062 - STAGED SURGICAL REPAIR FOR CHILDREN WITH INTERRUPTED AORTIC ARCH

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Background: Interrupted aortic arch is a rare congenital cardiac anomaly. Without treatment, mortality rate is approaching 90% within the first year of life.
Purpose: There are several surgical strategies for corrective repair of interrupted aortic arch. Here, we reported our surgical result of staged policy of corrective repair for the patients with interrupted aortic arch.
Method: From November 2003 to July 2015, there were 14 infants (8 boys, 6 girls) with interrupted aortic arch treated at our hospital. We routinely used intravenous infusion of prostaglandin E1 for all the infant patients to keep adequate end organ perfusion before the first surgical intervention. Then, staged surgery is our treatment policy.
Result: At the time of the first surgery, we did the first staged surgery with anastomosis between aortic arch and descending aorta, division of patent ductus arteriosus and banding of pulmonary trunk through left thoracotomy. The surgical survival rate of the first staged surgery was 100%. At the time of the second surgery, we did the corrective repair with the surgical procedures including repair of ventricular septal defect, de-banding of pulmonary trunk and enlargement of pulmonary trunk with pericardium under cardiopulmonary bypass through median sternotomy. The surgical survival rate of the corrective surgery was also 100%.
Conclusion: For the infants with interrupted aortic arch, ventricular septal defect and patent ductus arteriosus, we think the surgical result of staged repair can be approaching 100% by experienced surgeons and the good surgical result can be reproducible without significant difficulty.

P2063 - PRO BNP PROGNOSTIC VALUE IN PEDIATRIC PATIENTS WITH SINGLE VENTRICLE PHYSIOLOGY

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Purpose: To determine the prognostic value of pro-BNP level in patients with single ventricle physiology after hemodynamic corrections.
Methods: 70 children with physiology of single ventricle after hemodynamic corrections were included (January 2013 - December 2016). Age ranged from 6 days to 15 years. Standard clinical investigations, ECHO, 24-hours ECG-monitoring and pro-BNP levels were measured pre-operatively and 3, 6 months after. For analyze dates we have used Statisica 6.0 program.
Results: children from 0 to 3 years were predominant - 52 children (74%).Types of operation: BT-shunt– 10 patients (14.4%), Glenn procedure – 22 patients (31.4%), pulmonary artery bending – 14

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patients (20%), Norwood I surgery – 6 patients (8.5%), Fontan surgery – 18 patients (25.7%). Patients were divided into two groups. First group - patients with complications after surgery – 41 (58%); second group - patients without any complications – 29 patients (41.4%). The most common complications in the late post-operative period were heart failure (HF), arrhythmias. Mean pro-BNP levels in two groups pre-operatively were 1587 ± 52 pg/ml and 779 ± 34 pg/ml accordingly. In early post-operative period pro-BNP level was significantly higher in first group - 4576 ± 65 pg/ml and 998 ± 23 mg/ml, respectively ($p < 0.05$). After 3 months in the first group pro-BNP level was - 2942 ± 38 pg/ml, in the second group 389 ± 19 pg/ml ($p < 0.05$). 6 month after the first group still had high levels of pro-BNP, whereas in the second group pro-BNP level was near normal. Correlation analysis between pro-BNP level and morbidity in late post-operative period was performed. The value was 0.75.
Conclusions: Pro-BNP can be used as a morbidity prognostic marker in children after hemodynamic corrections, level $\geq$ 2500 pg/ml in early post-operative period predict development of arrhythmias.

P2074 - PATIENTS WITH AORTIC COARCTATION OPERATED WITHIN THE FIRST YEAR OF LIFE DIFFERENT SURGICAL TECHNIQUES AND PROGNOSTIC FACTORS 12 YEARS OF EXPERIENCE

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Background: The authors reviewed the clinical files of patients with coarctation of the aorta (CoAo) operated within the first year of life in two Italian centers, in order to assess the outcome of different surgical techniques and prognostic factors.
Materials and Methods: Patients operated between 2004 and 2016 were included in the study. Patients with complex heart defects (CHD) which underwent univentricular heart palliation were excluded. Several parameters were analyzed: sex, age at surgery, associated anomalies, associated surgical procedures, re-operation, mortality and need for re-intervention.
Results: 247 patients were operated during the study period; 61% were male. Surgery was performed at a median age of 13 days of life (range 0-346). 59% of the patients had isolated CoAo, 33% had CoAo associated to ventricular septal defect (VSD), and 7% had CoAo associated to other CHD. 85% of the patients were treated with end-to-end anastomosis, 12% with patch enlargement of the aorta, and 3% with a subclavian flap according to the presenting anatomy. During surgery 21% of the patients had an associated surgical procedure: banding of the pulmonary artery was performed in 70% of the cases, VSD closure in 15%, and arterial switch in 7.5%. One patient died after surgery due to sepsis; 17 patients were lost at follow-up. Among the remaining 229 patients, recoarctation occurred during follow-up in 24%. All but three underwent cardiac catheterization that was successful in 81% of the cases. Recurrence of coarctation was similar (22%) in patients who underwent either termino-terminal anastomosis or patch enlargement of the aorta, and it was higher in the group treated with subclavian flap (28%).
Conclusions: Mortality is very low. Risk of ricoarctation is almost 1/4 and is higher in patients who have been treated with subclavian flap. In case of recoarctation, cardiac catheterization is the first choice with good results.

P2075 - SURGICAL EXCLUSION OF DYSPLASTIC RIGHT VENTRICLE IN A CASE OF UHL'S ANOMALY

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We report a case a 2 year old boy with Uhl's anomaly presented with history of progressive respiratory distress, syncopal attacks and failure to thrive since birth. He underwent Surgical exclusion of right ventricle which included Bidirectional Glenn shunt anastomosis, atrial septectomy, interruption of main pulmonary artery, and fenestrated patch closure of tricuspid valve.Post-operative recovery was uneventful and anticoagulation was done. Biopsy report of right ventricle free wall showed only fibrous tissue with few blood vessels and nerve bundles. No myocardium was seen. There are very few successful surgical repairs in the literature and hence presenting our approach.

P2079 - UNIQUE CASE OF AORTICO LEFT VENTRICULAR TUNNEL (ALVT) ARISING FROM LEFT CORONARY NON CORONORY COMMISURE

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Aortico-left ventricular tunnel (ALVT) is a a paravalvular, extracardiac communication between aorta to left ventricle with a incidence of 0.001%.Most case reports shows it arising from right coronory sinus with very few reports shows arising from left coronory sinus. We report a unique case where it was arising from the commissure rather than sinus. Intraoperatively The aortic end of the tunnel was found to be around 15 mm X 15 mm was from the commissure with opening into both sinuses. The leftward end was close to the left coronary orifice with just enough margin for the sutures.The LV end was small, oval 8mm X 6mm below the base of non coronary cusp at aorto-mitral continuity..

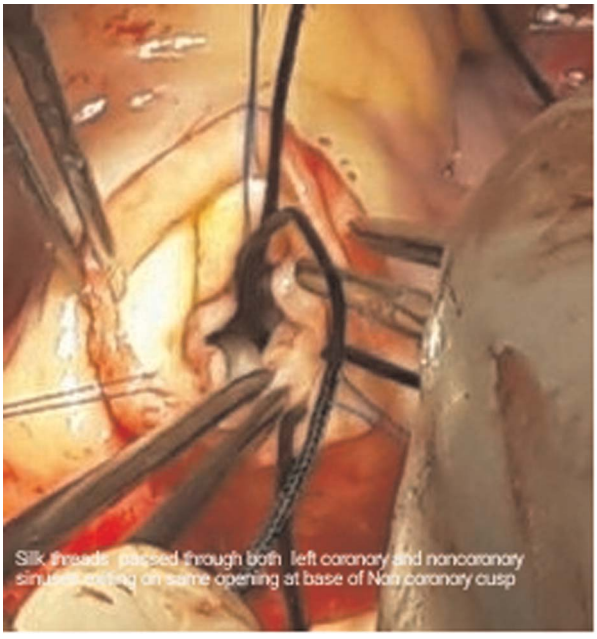

Figure 1.

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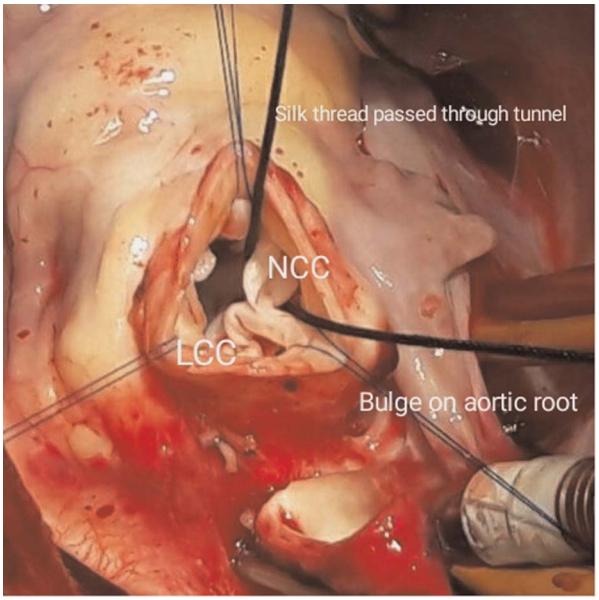

Figure 2.

P2080 - HIGH ALTITUDE MAY PROTECT AGAINST THE DEVELOPMENT OF EISENMENGER SYNDROME IN CHD PATIENTS

Alexandra Heath[1], Inge Von Alvensleben[1], Brian Graham[2], Rubin Tuder[2], Carlos Brockman[3], Erika Perez[3], Carla Arteaga[1], Claudia Scherer[1], Franz Fruedenthal[1]
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Background: Congenital heart disease (CHD) occurs at increased prevalence at high altitude, but there may be a later onset of the development of Eisenmenger syndrome. We hypothesized that CHD patients at high altitude are protected from the development of Eisenmenger syndrome.
Methods: Prospective observational case series study of patients who underwent surgery for CHD in La Paz, Bolivia, located at 3600 m. Patients aged 5 to 29 years (mean 12 years) with relevant post tricuspidal shunts (Qp/Qs 5,8 during hyperoxia test versus Qp/Qs :2,6 in hypoxia) underwent right heart catheterization pre-operatively and six to nine months post-operatively, and had a lung biopsy performed at the time of the surgery. Control lung tissue was obtained from patients who had pulmonary hydatid cyst resection.
Results: N = 10 CHD patients and N = 4 control patients were analyzed. Pre-operatively, the patients had significant response to hyperoxia with a fall in mPAP from 59,6 (DS 7,74) to 46,3 (DS11,1); (P = 0,05). Post-operatively, the patients had an excellent response to surgery, with a mPAP of 26,4 (DS6,42) (P < 0,05 versus pre-operative pulmonary pressures). Analysis of the lung histology did not show evidence of pulmonary vascular remodeling in the CHD patients compared to the control patients.
Conclusions: Chronic hypobaric hypoxia may protect CHD patients from early changes in the pulmonary vasculature and for instance from irreversible pulmonary hypertension at young ages.

P2094 - YOUNGER AGE AT FONTAN IS ASSOCIATED WITH BETTER 4.5 YEAR NEUROCOGNITIVE OUTCOMES

Billie-jean Martin[1], Gwen Bond[2], Joseph Atallah[2], Bryan Acton[3], Ari Joffe[4], M. Florencia Ricci[2], Gonzalo Garcia Guerra[4], Mohammed Al Aklabi[1], David Ross[1], Ivan Rebeyka[1], Charlene Robertson[2]
University of Alberta, Surgery, Edmonton-Canada[1]; University of Alberta, Pediatrics, Edmonton-Canada[2]; University of Saskatchewan, Pediatrics, Saskatoon-Canada[3]; University of Alberta, Critical Care, Edmonton-Canada[4]

Background: Optimal age for performing the Fontan operation is unclear, with recent suggestions that this step in single ventricle palliation should be delayed as long as possible to avoid accruing Fontan related complications. However, the association between age at Fontan and neurocognitive outcomes has not been previously assessed.
Methods: All subjects enrolled in the prospective Western Canadian Complex Pediatric Therapies Follow-up Program who underwent a Fontan operation between 1999 and 2015 were included. Follow-up clinical and neurocognitive data were obtained from the database. Psychologists completed neurocognitive testing on all surviving children at a median of 54 months of age using The Wechsler Preschool & Primary Scale of Intelligence. Generalized linear models were constructed to determine the association between pre-, intra- and post-operative variables, neuromotor or neurosensory disability, as well as characteristics over the course of palliation and neurocognitive outcomes.
Results: A total of 170 subjects (61 female, 97 with hypoplastic left heart syndrome) underwent a Fontan, with 163 surviving to undergo neurocognitive assessment. Median age at Fontan was 3.2 years (IQR 2.8, 3.7), and subjects most commonly had an extracardiac type Fontan (95, 56%). Twenty-two subjects had a neuromotor and/or neurosensory deficit diagnosed prior to undergoing a Fontan. Median Full-scale IQ (FSIQ) was 91 (IQR 77, 102), Performance IQ (PIQ) was 90 (78, 101), Verbal IQ (VIQ) 93 (81, 102) and Visual-Motor Integration (VMI) score 90 (IQR 78, 98). Older age at Fontan was associated with lower scores for each endpoint (Figure). In multivariable models including adjustment for any disability, age in years was a negative predictor for all neurocognitive endpoints (FSIQ -6.43; PIQ -6.60; VIQ -4.94; VMI -5.55; each p < 0.0001).
Conclusion: Younger age at Fontan is associated with better neurocognitive outcome at 4.5-years. The present trends towards delaying Fontan palliation needs to be considered in light of these findings..

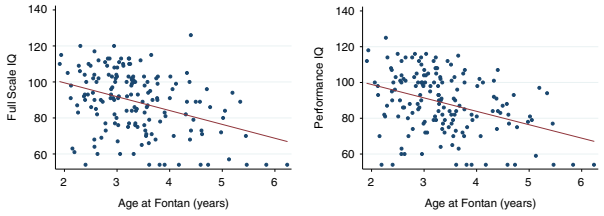

Figure.

P2097 - LEAVING A RIGHT VENTRICULAR PULMONARY ARTERY CONDUIT OPEN AT THE TIME OF BIDIRECTIONAL CAVOPULMONARY ANASTOMOSIS POST NORWOOD SANO OFFERS NO BENEFIT

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Introduction: It is unclear whether the Right Ventricular-Pulmonary Artery (RVPA) conduit should be left open at the time of bidirectional cavopulmonary anastomosis (BCPA) in Norwood-Sano patients, as the role of additional pulmonary blood flow in these patients post-BCPA has not been evaluated. We therefore aimed to evaluate the impact of additional pulmonary blood flow in this cohort.
Methods: All subjects that underwent a BCPA post-Norwood-Sano from 2002-2016 were included. Subjects were categorized according to the status of their RVPA post Norwood (OS – open Sano/PC- partially closed/CL – closed). Characteristics at the time of Norwood, BCPA and Fontan were compared across RVPA Status. The association between RVPA status and freedom from death and pulmonary artery intervention were assessed using cox proportional hazards models.
Results: 146 infants underwent a BCPA post-Norwood-Sano (53 female, 103 HLHS); 22 had their RVPA left OS, 56 PC, 68 CL. Subjects who had OS were more likely to have thrombosed head and neck veins pre-BCPA (45.4% vs 10.9% in PC vs 7.5% CL) and had significantly higher BCPA pressures (18.1 vs 16.9 PO vs 15.1 CL) and oxygen saturations post-BCPA (86% vs 84.2 PO vs 79.6% OS) (all $p < 0.05$). Patients with an open RVPA were younger at Fontan (3.2 OS vs 3.6 years CL, $p = 0.012$) and were more likely to suffer from post-Fontan stroke (OS 33.3%, PC 11.1%, CL 5.8%, $p = 0.042$). Furthermore, an open RVPA conduit was associated with increased mortality (OR 4.27 (1.29, 14.1)) but no reduction in need for pulmonary artery re-intervention (OR 0.57 (0.13, 2.51)).
Conclusions: Leaving the RVPA conduit open at the time of BCPA post-Norwood-Sano appears to offer no benefit but significant risk. This association may be due to the frequency of pre-BCPA venous thrombosis seen in the OS group, suggesting strategies to reduce thrombotic risk may also reduce myriad complications in this cohort.

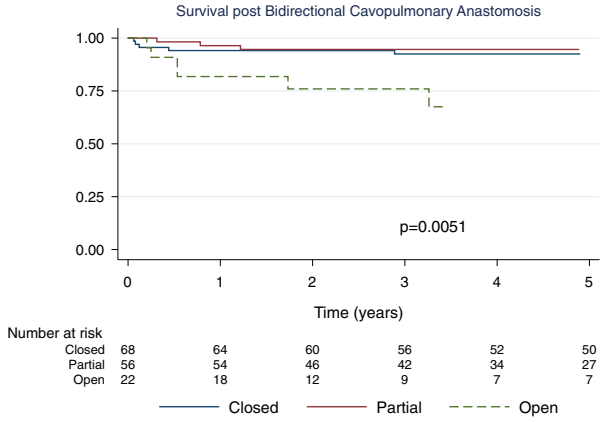

Figure.

P2108 - TAKEUCHI TECHNIQUE FOR TREE CASES OF ANOMALOUS LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY

Juan Miguel Mantilla[1], Jose Leonardo Cely-andrade[2], Jose Leonidas Olaya[3], Monica Marcela Moreno[3], Alfonso Velandia[4]
Hospital Cardiovascular Del Niño De Cundinamarca, Pediatric Cardiovascular Surgery, Soacha-Colombia[1]; Hospital Cardiovascular Del Niño De Cundinamarca - Universidad Del Bosque - Universidad Manuela Beltran, Research Department, Soacha - Bogotá-Colombia[2]; Hospital Cardiovascular Del Niño De Cundinamarca, Cardiovascular Surgery Departement, Soacha-Colombia[3]; Hospital Cardiovascular Del Niño De Cundinamarca, Cardiovascular Anhestesiology Departement, Soacha-[4]

Anomalous left coronary artery from the pulmonary artery (ALCAPA), also known as Bland-White-Garland syndrome (1) is a rare congenital cardiac disease that can cause myocardial infarction, heart failure and even death in paediatric patients (2), is present in 1 of 300 000 live births and accounts for 0.05% of congenital cardiac malformations (3). Surgical treatment is ever the treatment election and success overlive is most than 90%; coronary reimplatation is now the elective treatment when it is not possible, takeuchi technique is the elective treatment; this technique repairs intrapulmonary tunnel and is a unique procedure for repairing anomalous origin of the left coronary artery with left and posterior possition from the pulmonary artery (3) Four female clinical cases are presented: a 4 years old, 5,8 y10-months all patients with dilated cardiomyopathy with severe ventricular dysfunction, LVEF 15%, 20% 20% and and 40%, respectively, with moderate mitral insufficiency secondary to anomalous origin of the left coronary artery Of the Pulmonary Artery Trunk (ALCAPA), Takeuchi surgery was performed in all cases, with excellent results given by recovery of ventricular function as a whole. Follow-up time of 6 to 3 years.

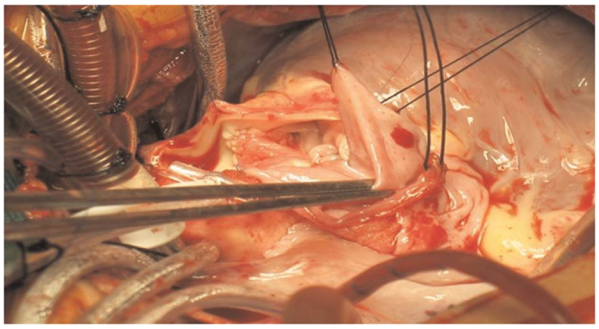

Figure 1.

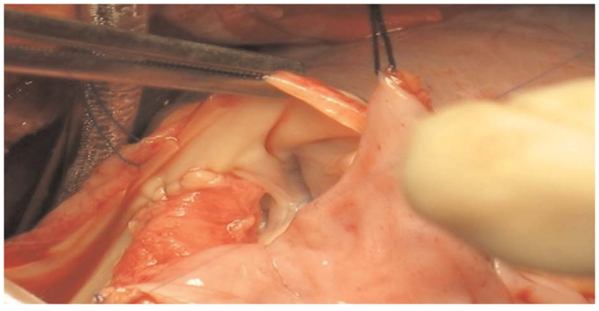

Figure 2.

Table 1. Characteristics of patients who underwent surgical repair with takeuchi technique.

Year	Age	Pre surgical LVEF	Post-surgical LVEF	Follow up LVEF (time)	Followup MI
2008	4 years old	15%	38%	65%65% (3 years)	Moderate
2013	8 months	20%	45%	65%60-70% (3 years)	Moderate
2015	5 months	20%	45%	65% (1 year)	Moderate
2016	9 months	40%	40%	65% (6 months)	Low

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P2113 - WHICH IS THE EXACT SIZE OF THE TRANSANNULAR PATCH NEEDED TO ACHIEVE A NORMAL DIAMETER OF THE PULMONARY ANNULUS DURING SURGICAL CORRECTION OF TETRALOGY OF FALLOT

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Introduction: The main concern about using a transanular patch (TAP) during surgical correction of tetralogy of Fallot is that insufficient enlargement of the annulus leaves residual stenosis and excessive enlargement aggravates pulmonary regurgitation (PR). PR impairs right ventricular function and increases postoperative mortality. Thus, achieving a normal postoperative pulmonary annulus diameter (PAD) becomes highly desirable.
Materials and Methods: From March to December 2016, 20 patients with hypoplastic pulmonary annulus (defined as PAD < -2 z-value) were operated on in the two involved institutions. Patients: 9 males and 11 females; median age: 13 months; mean weight: 9.4 kg (6.3–16). Transthoracic echocardiography was performed to determine the preoperative PAD (mean: 6.85 mm; range: 6–10). Exact width of the TAP for each possible situation was obtained by subtracting the perimeter of the preoperative pulmonary annulus from the normal one. Values are depicted in Table 1.
During surgery, the right ventricular outflow tract, the pulmonary annulus and the main pulmonary artery were enlarged using a glutaraldehyde-treated autologous pericardial patch cut to size according to this geometrical rule.
Results: There were no postoperative deaths. PAD measured by transthoracic echocardiography in the early postoperative period ranged from 0.9 mm smaller to 1 mm larger than respective normal value. Mean postoperative PAD was 11 mm for a mean normal annulus of 11.1 mm. Mean percentage of deviation of the post-operative PADs from respective normal values was 0.6%. Average transpulmonary gradient was 26 mmHg, meaning mild residual pulmonary stenosis. PR was moderate in 50% of the cases and mild or less in the remaining patients.
Conclusion: This geometrical rule accurately defines the exact width of the TAP needed to achieve a normal postoperative PAD, avoiding residual gradients and severe PR in order to prevent right ventricular dilation in the follow-up of these patients.

Table.

Patient's PAD (mm)	**1**	**2**	**3**	**4**	**5**	**8**	**7**	**8**	**9**	**10**	**11**	**12**	**13**	**14**	**15**
Weight (kg)															
2.9	23	20	18	13	10										
4.1	28	23	20	17	14	11									
5.4	29	26	23	20	16	13	10								
6 7	32	28	25	22	19	16	13								
8	33	30	27	24	21	18	15	11							
9.4	35	32	29	26	23	20	16	13	10						
10.8	37	34	31	27	24	21	18	15	12						
12.3	38	35	32	29	28	23	20	18	13	10					
13.8	40	37	33	30	27	24	21	18	15	11					
15.3	41	38	35	32	28	25	22	19	18	13					
18.9	42	39	38	33	30	27	23	20	17	14	11				
18.8	43	40	37	34	31	27	24	21	18	15	12				
20.3	44	41	38	35	32	28	25	22	19	18	13				
22	45	42	39	38	33	29	28	23	20	17	14	11			
23.8	48	43	40	37	33	30	27	24	21	18	15	11			
25.7	47	44	41	38	34	31	28	25	22	19	18	12			
27.8	48	44	41	38	35	32	29	28	22	19	18	13	10		
31.7	49	48	43	40	37	33	30	27	24	21	18	15	11		
38	50	47	44	41	38	35	32	28	25	22	19	18	13		
40.7	52	49	45	42	39	38	33	30	27	23	20	17	14	11	
45.7	53	50	47	43	40	37	34	31	28	25	22	18	15	12	
51.2	54	51	48	44	41	38	35	32	29	28	22	19	18	13	10
57	55	52	49	48	43	39	38	33	30	27	24	21	17	14	11
63.4	58	53	50	47	43	40	37	34	31	28	25	22	18	15	12
70.4	57	54	51	48	44	41	38	35	32	29	28	22	19	18	13

PAD: pulmonary annulusdiameter

P2119 - COMPARISON OF MINIMALLY INVASIVE TRANSTHORACIC DEFECT CLOSURE AND CARDIOPULMONARY BYPASS FOR THE TREATMENT OF SUBARTERIAL VENTRICULAR SEPTAL DEFECT IN PEDIATRIC PATIENTS

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The Children, The Heart Center, Hangzhou-China[1]; The Children's Hospital of Zhejiang University School of Medcine, The Heart Center, Hangzhou-China[2]

Objective: To investigate the feasibility, safety and effectiveness of MITDC for the treatment of subarterial VSD.
Methods: Analyzed clinical data of pediatric patients diagnosed of subarterial VSD by echocardiography from October 2011 to September 2016. There were a total of 54 cases underwent MITDC and 91 cases received surgery under CPB. Patient's clinical data before and after surgery were collected, and efficacies of the two methods were compared.
Results: There was no significant difference in age, gender and body weight between the two group of patients ($P > 0.05$);defect diameter of the MITDC group was significantly smaller than that of the CPB group ($P < 0.01$), and the percentage of right aortic sinus prolapse in the MITDC group was lower than that in the CPB group ($P < 0.01$). Postoperative ICU intubation time and duration of hospital stay of the MITDC group were both shorter than that of the CPB group ($P < 0.01$), and the percentage of patients requiring blood transfusion was significantly lower in the MITDC group than in the CPB group ($P < 0.01$). Compared with the preoperative level, rate of mild postoperative tricuspid regurgitation among patients in the MITDC group was significantly reduced ($P < 0.01$). The MITDC group had a significantly lower postoperative pulmonary peak flow velocity than the CBP group ($P < 0.01$). The percentage of postoperative incomplete right bundle branch block was higher than that the preoperative rate in both groups ($P < 0.01$), the rate of postoperative arrhythmia remained the same as compared to the preoperative level in both groups ($P > 0.05$). There was no statistically significant difference in the rate of pericardial effusion between the MITDC and the CPB groups ($P > 0.05$). There was no other complication observed in the two groups except the above mentioned complications.
Conclusion: MITDC is an effective procedure for the treatment of subarterial VSD, MITDC had the advantages of less trauma, shorter ICU intubation time, shorter hospital stay, less need for blood transfusion.

P2121 - NEW VALVE SPARING TECHNIQUE FOR TETRALOGY OF FALLOT REPAIR

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Amrita Institute of Medical Sciences And Research Centre, Pediatric Cardiac Surgery, Kochi-India[1]; Amrita Institute of Medical Sciences and

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Research Centre, Cardiac Anesthesia, Kochi-India[2]; Amrita Institute of Medical Sciences and Research Centre, Pediatric Cardiology, Kochi-India[3]

Background: We present a modified technique of valve sparing surgery to preserve the pulmonary annulus and prevent pulmonary regurgitation (PR) following intracardiac repair (ICR) for Tetralogy of Fallot (TOF).
Methods and Results: The data of 86 consecutive patients (median age 11 (3-161) months) undergoing TOF repair (March 2015-December 2016) was reviewed. Transannular patch (TAP) was avoided in 55 (64%); 25 (29%) required only infundibular resection and pulmonary valvotomy (trans-atrial ICR) while 30 (35%) patients underwent modified valve sparing surgery (mVS-ICR) to preserve the pulmonary valve (PV) and pulmonary artery (PA) annulus using following techniques: commissurotomy, commissural release, sinus incisions in the PA, T-incision of right ventricular out flow tract (RVOT) and balloon dilatation of PA annulus, leaflet delamination to increase leaflet height and achieve coaptation in the setting of the new larger PA annulus and, augmentation of individual sinuses and RVOT with autologous treated pericardium. There was no difference in the age, weight, surgical support times, intensive care and ventilation duration and mortality between the three groups. Key results are summarized in the table.
Conclusion: The modified valve sparing correction of TOF can be used to preserve the valve in selected patients with small PA annulus. Short-term results appear promising. Long-term follow-up is required to determine growth of PA annulus and progression of PR..

Table 1. Results of modified valve sparing ICR (mVS-ICR) shown in comparison with the Trans-atrial ICR and transannular patch (TAP) groups

	Number	PA Annulus Z-Score mean (SD)	Main Pulmonary artery Z- score mean (SD)	In-hospital mortality	Follow up (median 9 months (range 1-22)	
					PR	RVOTO
mVS-ICR	30 (35%)	-2.54 (1.02)	-4.06 (1.34)	2	2 (7.1%)	3 (10.7%)*
Trans-artial repair	25 (29%)	-0.5 (0.77)	-1.24 (1.26)	0	1 (4%)	0
TAP	31 (36%)	-3.46 (1.36)	-3.98 (1.97)	1	26 (86.7%)	2 (6.9%)

* gradients of 40, 56 and 36

P2122 - SURGICAL EXCISION OF CARDIAC TUMORS 20 YEAR EXPERIENCE AT A SINGLE INSTITUTION

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Alder Hey Children Hospital, Cardiac Surgery, Liverpool-United Kingdom

Background: Cardiac tumors are rare with heterogeneous histology and location. Indication for surgical resection is dictated by symptoms and position. The aim of the present study is to review our institution's results in patients who underwent resection of cardiac tumors since 1997.
Materials and Methods: Retrospective review from January 1997 to January 2017 was accomplished. We followed up all patients including echocardiography assessing regression or progression.
Results: 13 patients with a diagnosis of cardiac tumor were operated at our Institution in the study period. The demographic of the population and the histology subtype are described in the Table. -Rhabdomyomas: Four of the six patients were operated in neonatal age, the remaining two at 2 and 3 months of age. In 4 of the 6 patients, the tumor was in the LVOT causing significant obstruction, while in two patient2 there was RVOT obstruction. -Myxomas: Patients with this subtype were older. All the myxomas were excised from the left atrial septum repaired with a patch. One patient had associated mitral valve repair. -Pericardial Teratoma: One patient was operated in neonatal age, while the second was 10 months old. One patient with extension of the tumor into the aortic root died 23 days postoperatively for massive haemorrhage following aortic root repair. -Fibroma: one patient had a fibroma excised from the RV free wall at 8 months of age -Haemangioma: one patient with right atrial haemangioma encasing the RCA was successfully operated and the coronary repaired. We had one (1/13 7.6%) in hospital death and no late death. At a mean follow up time of 77 + /- 67 months no patient required reoperation and echocardiogram excluded regrowth of the tumour.
Conclusions: Surgical excision can be performed with low mortality even when patients are operated in neonatal age. We showed good long term follow up with no regrowth of tumor.

Table.

	N°	Gender M-F	Median Age (Range)	Median Weight (Range)	Position
Rhabdomyoma (1RV + IVS), 1 RVOT, 1 RVOT and IVS	6	0-6	11d (3d-82d)	3 Kg (2.4-4.6)	4 LVOT
Myxoma	3	2-1	4.5yrs (46 m-138 m)	21 Kg (19-25)	2 LA + IAS 1 LA
Pericardial teratoma	2	1-1	5 m (3d -10 m)	5.25 Kg (2.4-8.1)	1 pericardium 1 pericardium/ Ao. root
Fibroma	1	1-0	6 m	6.38 Kg	1 RV free wall
Haemangioma	1	1-0	27d	3.7 Kg	1 RA

LVOT=left ventricle outflow tract; RVOT=right ventricle outflow tract; IVS=interventricular septum; LA = left atrium; RV = right ventricle; RA = right atrium; Ao. = aortic d = days; m = months; yrs = years

P2130 - LIMITED EXTREME LATERAL CAVOATRIOTOMY FOR SURGICAL CORRECTION OF ANOMALOUS PULMONARY VENOUS CONNECTION FACILITATED BY LIMITED POSTERIOR THORACOTOMY APPROACH

Luis Baquero[1], *Brijesh Kottayil[1], Praveen Reddy[1], Amitabh Sen[2], Sumangala Sunil[1], Krishna Kumar[3]*
Amrita Institute of Medical Sciences, CVTS, Kochi-India[1]; Amrita Institute of Medical Sciences, Cardiac Anesthesia, Koch-India[2]; Amrita Institute of Medical Sciences, Pediatric Cardiology, Kochi-India[3]

Introduction: Surgical correction of partial anomalous pulmonary venous connection (PAPVC) to Superior venacava (SVC) is often associated postoperatively with Sinus node dysfunction and venous obstruction. We present a modified approach by limited right posterior thoracotomy (LPT) for surgical correction of this anomaly which has excellent cosmetic advantage and also facilitates surgical correction by our modified extreme lateral cavoatriotomy approach which avoids injury to the sinus node.
Methods: 53 patients who underwent surgical correction of PAPVC to SVC with our modified approach between January

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2010 and December 2016 were evaluated retrospectively. All patients underwent surgical correction by LPT with median extent of the incision stopping short of posterior axillary fold. Modified extreme lateral cavoatriotomy incision is made flush with the draining pulmonary vein facilitated by thoracotomy approach which brings the draining pulmonary veins parallel to the surgical view and atrial incision stops short of cavoatrial junction avoiding injury to sinus node.
Results: Median age was 4 years (1-25 years). Median weight was15 kg (6-63 kg). At a median follow up period of 27 months (1 -60 months) there were no early or late mortality. None of the patients had any sinus node dysfunction or arrhythmia.On follow up there were no significant SVC or pulmonary venous obstruction. One patient had mild flow acceleration in SVC with mean gradient of 3 mm of hg. All patients had excellent wound healing with cosmetic advantage over standard median sternotomy approach.
Conclusion: Extreme lateral cavoatriotomy approach via LPT is a safer technique for surgical repair of PAPVC to SVC with excellent cosmetic advantage. In addition this technique potentially offers the patient the lowest risk for the development of sinus node dysfunction after surgical correction.

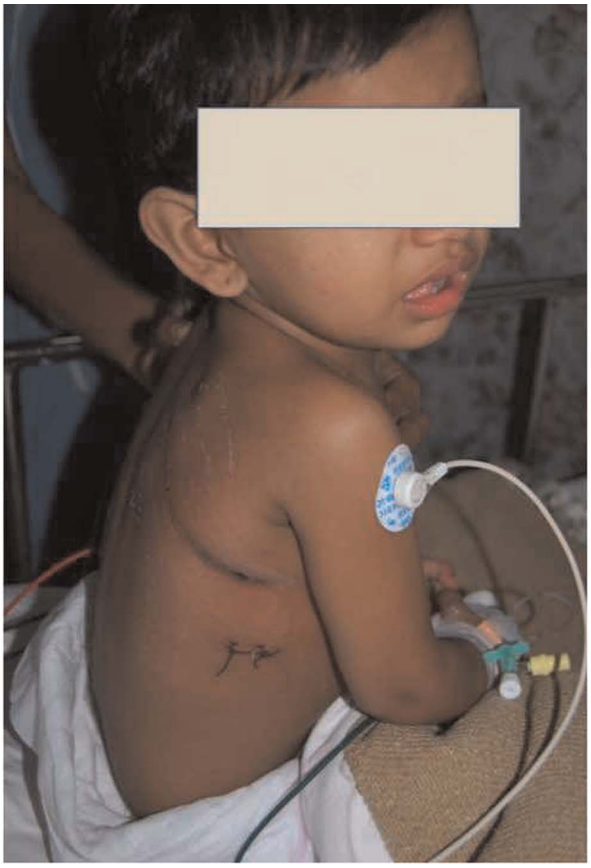

Figure.

P2134 - COMPREHENSIVE SURGICAL APPROACH DURING REPAIR OF MULTIPLE MUSCULAR VENTRICULAR SEPTAL DEFECTS

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Background: To eliminate the left to right shunt completely during intra cardiac repair of multiple VSD is a surgical challenge. We adopted a comprehensive surgical approach to minimize the residual shunt after repair.
Methods: Between March 2012 to December 2016 we did primary repair of multiple VSD in 52 patients. The age ranged from 3 months to 12 years (median 8 months). The weight ranged from 2.7 to 11 kg (median 4.6 kg). Intraoperatively, we closed the major VSD's with separate pericardial patches (70 VSD's). The moderate and smaller sized anterior muscular, mid muscular and apical VSD's were addressed by endothelial suturing using continuous polypropylene suture in two layers tied on pericardial pledgets (37 patients). The apical and mid-muscular VSD's which had a single opening on the LV side and sizeable multiple holes on the RV side were addressed by double breasting using teflon patches on both sides of the septum and suturing the RV side of the patch to the endocardium by continuous suturing (8 patients).
Results: Intraoperatively, a bubblegram with TEE guidance was performed using the LV vent in addition to SVC and PA saturation step up. There was no operative mortality. Three patients required re-operation for significant residual shunt in the immediate post-operative period. Two patients died in the late post-operative period due to lung infection. All the other patients have been followed up (median follow up 22 months) and are doing well without congestive heart failure. 2D echo evaluation during follow up showed no significant residual shunts.
Conclusions: A combination of surgical techniques is required to manage multiple VSD's. Endothelial suturing is a very useful surgical strategy and preserves LV function without compromising RV cavity. Double breasting with teflon patches should be reserved for selective cases.

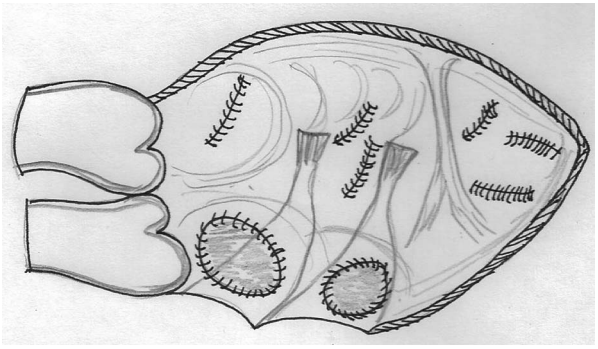

Figure.

P2136 - IMPACT OF DELAYED STERNAL CLOSURE FOLLOWING PAEDIATRIC CARDIAC SURGERY ON WOUND INFECTIONS

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Background: Delayed sternal closure (DSC) is a well-established procedure to reduce postoperative hemodynamic and respiratory instability following neonatal or infant cardiac surgery. Though, DSC is associated with an increased rate of sternal wound infection

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(SWI), sepsis and mortality. Currently, indication and duration of DSC vary substantially between paediatric cardiac centres worldwide. Therefore, we reviewed our experience with DSC to determine the incidence of SWI and to evaluate potential perioperative risk factors.
Materials and Methods: A retrospective study was conducted of 55 consecutive paediatric patients (33 male, 22 female; median age 14 days, range 1 day - 14 years; median body weight 3.6 kg, range 2.0–63.0 kg) who underwent DSC between January 2013 and December 2014 at our centre. DSC was routinely performed after initial palliation for single ventricle disease (29%; n = 16). In all other cases decision for DSC was made electively inside the OR.
Results: Median duration of sternal opening was 3 days (1 to 32 days). Overall mortality was 14.5% (n = 8). Eight patients experienced SWI following DSC and wound cultures were positive in six cases, detecting staphylococcus aureus (n = 3), coagulase-negative staphylococcus (n = 2), and staphylococcus epidermidis (n = 1). One patient needed surgical revision for a deep SWI four weeks after chest closure. An indipendent perioperative risk factor for SWI was former support with ECMO (OR 7.07, 95% CI 1.27-39.41, p = .03). There was no significant correlation between SWI and prolonged cardiopulmonary bypass time (>180 min), aortic clamp time (>100 min), length of ICU stay (>7 days), duration of open chest (>4 days) and usage of foreign material during operation.
Conclusion: DSC is a safe and feasible procedure to avoid postoperative hemodynamic instability. Although, sternal wound infection is an aggravating side effect of DSC, especially in patients with ECMO, surgical revision is rarely needed.

P2137 - COMPREHENSIVE SURGICAL APPROACH DURING REPAIR OF MITRAL VALVE IN CHILDREN WITH SPECIAL EMPHASIS ON POLYTETRAFLUOROETHYLENE NEO CHORDAE

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Background: Repair of congenital mitral valve disease is a challenge. A combination of techniques are required to and avoid valve replacement which is a very difficult option in this subset.
Methods: Between June 2013 to December 2016, 30 patients underwent repair for mitral valve disease. The age ranged from 2 months to 12 years (median 24 months). The weight ranged from 3.7 to 22 kg (median 8.5 kg). Six patients had mitral stenosis (3 sub-valvular fusion and 3 supra-valvular ring). The remaining patients had mitral regurgitation and among them, the associated lesions were VSD (12), ASD (5) and ALCAPA (1). Three patients had an associated tricuspid valve repair. In patients with stenosis, resection of the ring and splitting of the sub-valvular papillary muscles was done. 15 patients had significant prolapse of the AML requiring caudal shortening in 2 and insertion of polytetrafluoroethylene (PTFE) neo-chordae in 13. In ALCAPA, quadrangular resection of the AML was done to reduce the bulk before insertion of the neo-chordae. In 3 patients, a separate scallop was repaired. For annular dilatation, a posterior annuloplasty was done with pericardial strip in 14 and a commissuroplasty in 5.
Results: There was no operative mortality. There was one late mortality in the 2 month old infant due to respiratory infection. During median follow up of 16 months, all the remaining patients were doing well with no significant decongestive therapy. Mitral regurgitation assessed was moderate in 8 patients and mild in the remaining 15 patients.
Conclusions: A combination of techniques are required during repair of mitral valve in children. Use of PTFE neo-chordae is always helpful to reduce prolapse and regurgitation. Avoiding valve replacement is an important strategy, but however long-term follow up is required to see if moderate regurgitation progresses.

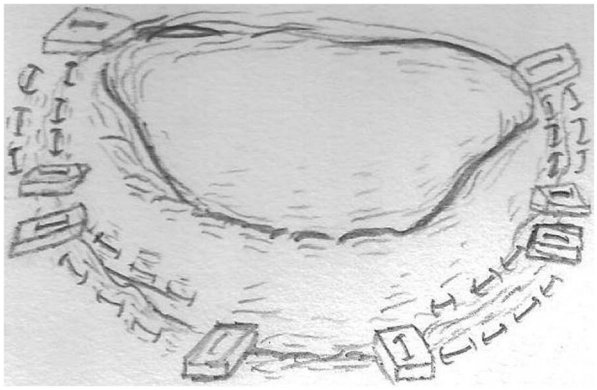

Figure 1.

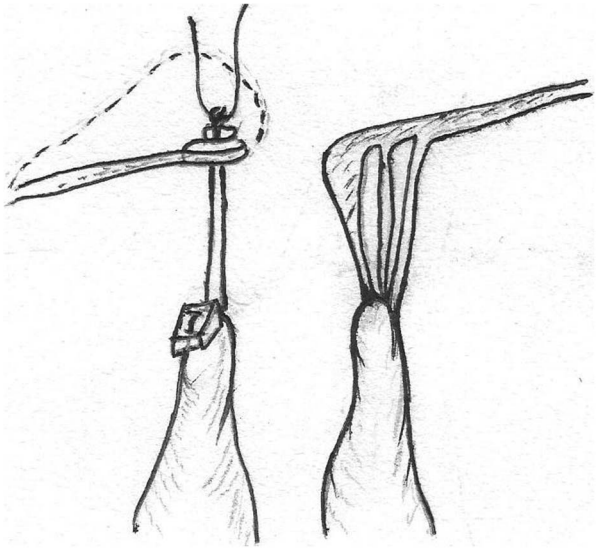

Figure 2.

P2138 - A NOVEL REPAIR TECHNIQUE OF CONGENITAL ABSENCE OF RIGHT PULMONARY ARTERY

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Objectives: Isolated unilateral absence of a pulmonary artery (UAPA) is a rare lesion and early surgical repair of UAPA deems necessary to maintain pulmonary blood supply. Surgical approaches include creation of an aorto-pulmonary shunt or connection of a MAPCA, if present, to the main pulmonary artery (MPA) or

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aorta. Here, we present a novel surgical technique using an autologous interposition tube graft, created from the roof of the MPA, to bridge the discontinued RPA.
Methods: An 8-days-old female (bodyweight 2,8 kg; 48 cm) with respiratory distress (SaO2 ~ 85%) was referred to our centre. Echocardiography revealed an absent RPA without other cardiac malformations. Catheterization depicted a normal-sized LPA and a ductus diverticulum at the brachiocephalic artery indicating a closed right ductus arteriosus originating from the hilar RPA (Figure1A). At surgery, a 1,5 × 3 cm piece of tissue was resected from the anterior wall of the MPA. To create a "tube", the graft was sutured around a 3mm Hegar pin. To get better access to the new RPA "bed", the ascending aorta was transected. Next, the "tube" was anastomosed to the hilar RPA and to the MPA. The MPA roof was augmented with an autologous pericardial patch (Figure1B-D).
Results: Follow-up was complicated by an anastomotic stenosis on the level of the hilar RPA. Therefore repeated balloon dilatation (Savvy 3mm/20 cm; SeQuent Please 4.0 × 17mm; BARD ultraverse 5 × 20mm) and finally stent implantation (Formula Stent 8 × 20mm; 7th postoperative month) were performed with good angiographic results (Figure1E). Today, 9 months after the inital operation, the RPA has an acceptable diameter (4mm) and the patient is thriving very well.
Conclusion: We describe a novel pulmonary graft interposition technique for UAPA repair using complete autologous material. Hence, the chance of future growth of the new RPA is increased with our approach. However, graft stenosis is of concern, mandating close follow-up and tailored treatment.

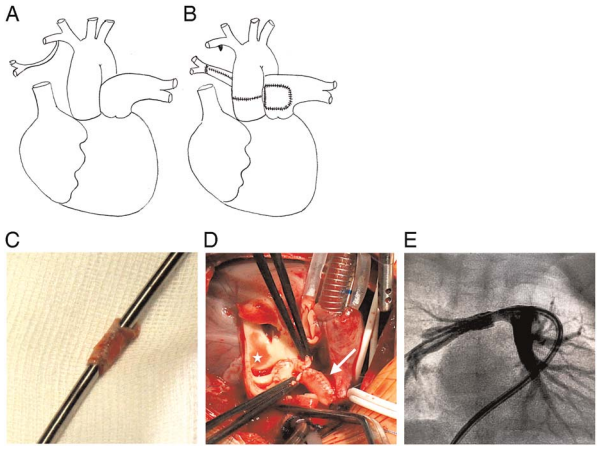

Figure 1.
Preoperative cardiac anatomy - absence of the right pulmonary artery, persistence of a right ductus arteriosus connecting the brachiocephalic artery with the hilar RPA (A). Postoperative cardiac anatomy - "tube" graft now in RPA position, MPA roof augmentation with autologous pericardial patch, transected aorta ascendens, excised right ductus arteriosus (B). Intraoperative creation of a "tube" graft using a 3mm Hegar pin 3mm and PDS 7x0 (C). Anastomosis of "tube" graft (white arrow) to the hilar RPA and MPA (white star) (D). Right heart catheterization after RPA stenting shows an acceptable RPA diameter (E).

P2140 - POST CARDIOTOMY EXTRACORPOREAL MEMBRANE OXYGENATION IN NEONATES – BENEFITS OF DELAYED SYSTEMIC HEPARINISATION

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Background: Extracorporeal membrane oxygenation (ECMO) is a well-established treatment option for infants with postcardiotomy heart failure. However, ECMO support is associated with major complications such as excessive bleeding and thrombosis. Even though, a standardized protocol for anticoagulation on ECMO is currently missing. Thus, we seek to report our experience with a specific anticoagulation regime in neonates with postcardiotomy ECMO. The purpose of the present study was to investigate the impact of delayed systemic heparinisation on thrombosis (1), bleeding (2) and requirement of blood products (3) during ECMO treatment.
Materials and Methods: Retrospectively, we reviewed medical charts of fifteen neonates (mean age 13 ± 2.6 days; mean weight 3.1 ± 0.3 kg) who were placed on ECMO after biventricular or palliative cardiac surgery at our centre between October 2012 and May 2014. In all patients, anticoagulant effects of heparin were reversed by administration of protamine, platelets and fibrinogen (target ACT 120 ± 20 sec) with the beginning of ECMO support. Systemic heparinisation was delayed for 18 hours (range 8 – 37).
Results: Mean duration of ECMO support was 4.5 days. During that period, no thrombotic events were observed. 3 patients (20%) required re-thoracotomy for bleeding on the first postoperative day. Transfusion volumes were 24.5 ml/kg/d of packed red blood cells, 9.6 ml/kg/d of fresh frozen plasma and 7.5 ml/kg/d of platelets. Survival to hospital discharge was 87%. During follow-up (mean 15.5 month) there were no late thrombotic events observed.
Conclusion: Neonates with post-cardiotomy ECMO benefit from delayed systemic heparinisation in terms of low risk for thrombosis, reduced bleeding rates and low transfusion volumes. Though, research is clearly limited in this field. To improve neonatal and paediatric outcome on ECMO, a standardized anticoagulation protocol is urgently needed.

P2147 - SEVERE COMPLICATIONS OF INFECTIVE ENDOCARDITIS (IE) REPORT OF TWO CASES

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Background: IE has nonspecific symptoms, so early diagnosis is often difficult. It presents a high morbidity and mortality. Complications such as abscesses and aneurysms are rare but potentially lethal. Both had poor dental hygiene.
Case 1: Nine-year-old boy with bicuspid aortic valve previously submitted to valve replacement. He was admitted with a history of fever for two weeks, weakness and conjunctival petechiae. Echocardiogram showed vegetation on the atrial side of the anterior leaflet of the mitral leaflet, which was also perforated and supravalvar aortic vegetation. Blood culture was positive for S. viridans and he was started on antibiotics. Computed tomography (CT) showed sings of hepatic, splenic and cerebral embolism. He was submitted to an emergency surgery, when an abscess was found in the fibrous skeleton of the heart and ascending aorta, as well as vegetations in the mitral and aortic valves. Successful Konno-Bentell surgery was performed. The histopathological

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study demonstrated active IE. He had six weeks of venous antibiotic treatment after surgery, with good clinical outcome.
Case 2: Six-year-old boy, with previous diagnosis of ventricular septal defect (VSD), right ventricular muscle band and aortic regurgitation, with fever for four weeks and hyporexia. Echocardiogram showed a peduncular vegetation in the mitroaortic region and an aneurysm that extended from the base of the anterior mitral leaflet to the aortic root, measuring around 12 mm. He was submitted to surgery, with removal of vegetations, closure of the aneurysm ostium, aortic valve repair and ventriculoseptoplasty. Antibiotic therapy was continued for four weeks after surgery, with good clinical outcome.
Discussion: IE is a disease that should always be considered as a differential diagnosis of persistent fever, especially in children with structural disease and poor dental hygiene. The two reported cases of complicated IE had a favorable outcome, despite the severity of the lesions.

P2148 - PREDICTORS OF SURGICAL OUTCOME OF CRITICAL CONGENITAL HEART DISEASE IN THE DEVELOPING WORLD

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Background: Despite a major improvement of cardiac surgery in neonates and infants with critical congenital heart disease (CCHD) currently, the mortality and morbidity of these patients remain high. Recent data of risk factors and surgical outcome of CCHD had been scanty reported among developing countries.
Objectives: To describe the patient characteristics and surgical procedures and to identify risk factors associated with in-hospital mortality in infants with CCHD at a single tertiary care center.
Method: Retrospective study of CCHD in infants who were newly diagnosed and underwent cardiac operation at Chiang Mai University Hospital, Thailand from January 2011 to December 2015.
Results: Among 183 critical congenital heart disease infants, 58% were male and the median gestational age was 38 weeks (range 27-42 weeks). The average birth weight was 2,873 ± 537 grams and 22 infants (12%) were preterm. The most common cardiac defect was TOF/PA or severe TOF (18.5%), followed by complete TGA (17%), COA or IAA (15%), single ventricle (14%), DORV (11%) and others (24.5%). Surgical procedures included modified BT shunt or central shunt (39%), simple or complexed COA or IAA repair (21%), arterial switch (15%), PA banding (9%), TAPVR repair (6%) and others (10%). The median age at palliative or corrective cardiac surgery was 29 days (interquartile range 12-89). The in-hospital mortality was 16.4% (30 of 183). Preoperative factors associated with in-hospital death were prematurity (OR 4.23, 95% CI 1.08-16.53, p = 0.038), and prenatal diagnosis (OR 6.45, 95% CI 1.59-26.18, p = 0.009), and postoperative risk factors for mortality were ECMO insertion (OR 14.23, 95% CI2.99-68.19, p = 0.001) and renal failure (OR 7.23, 95% CI 1.78-29.83, p = 0.006).
Conclusion: Surgical mortality for infants with critical congenital heart disease remains high in developing world with limited resource. Preterm infants were associated with high in-hospital mortality.

P2152 - ACUTE DYSFUNCTION OF RIGHT VENTRICULAR OUTFLOW TRACT VALVED CONDUIT IN THREE CASES IN THE FORM OF ANEURYSMAL DILATATION AND THROMBOSIS

Okan Yildiz[1], Erkut Ozturk[2], Behzat Tuzun[1], Bahadir Genc[1], Selman Gokalp[2], Alper Guzeltas[2], Sertac Haydin[1]
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Background: The Contegra® bovine jugular venous conduit is used to maintain continuity between the right ventricle and the pulmonary arteries. Younger patients and smaller grafts are risk factors for Contegra® failure and complications are possible due to variable flow in the distal pulmonary vasculature. However, early (first 30 days) aneurysmal dilatation and thrombosis are not common and have not been reported as a series. We would like to present 3 cases related to Contegra® dysfunction and replacement in early period.
Case Report: Between November 2014 and November 2016, 17 pediatric patients underwent right ventricular out flow tract reconstruction with Contegra® valved conduits. Three of these patients (<6 months) developed Contegra® (size <14 mm)

Table 1. Patients'demographics

	Case 1	Case 2	Case 3
Age/Weight (kg)	3 months, 5.2	5.5 months, 5	3 months, 5.1
Diagnosis	TGAyVSD/severe PS with annular hypoplasia	PAyVSD/MAPCAs	Truncus arteriosus
Distal pulmonary vasculature	Hypoplastic PAs Mcgoon index; 1,7	HypolasticPAs	Normal PAs PHT
Surgery	Rastelli operation	Rastelli operation	Truncus repair
Si ze of Contegra® (mm)	12	14	12
Concomitant procedures	no	MAPCA unifocalization and pulmonary artery plasty	Right coronary artery osteoplasty
CPB, minutes	120	114	155
Cross clam pi ngti me, minutes	92	77	122
RV/LV pressure at the end of the operation	0,45	0,6	0,45
ECLS start time (hours)	24	12	18
Indi cation for ECLS	E-CPR	RVfailure-LCOS	RVfailure-LCOS
ECLStime(days)	7	3	11
Cardiac cathtime, postoperative (days)	5	3	4
Revision time, postoperative (days)	5	3	4
Si ze of Aneurysm atic Contegra® (mm)	16	21	19
Thrombus	Acute thrombus, coveringthe valve cusps andthe conduit itself	Macroscopical1y, thrombus coveringthe valve cusps	Macroscopically, thrombus coveringthe valve cusps
Conduit regurgitation	No valve movement	Severe	Severe
Mechanism of Conduit dysfunction	Thrombosis, Aneurysmal Dilatation	Thrombosis, Aneurysmal Dilatation, insufficiency	Thrombosis, Aneurysmal Dilatation, insufficiency
Replaced With	11 mm Mathx-Pplus N® decellularized porcine pulmonary conduit.	12 mm Contegra® bovine jugular venous conduit.	11 mm Labcor® stentless porcine valved pulmonary conduit

CPB; Cardiopulmonary bypass, LCOS; Low Cardiac Output; ECLS; extracorporeal life support; E-CPR, extracorporeal cardiopulmonary resuscitation; PAs; Pulmonary Arteries

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dysfunction in the early postoperative period (details are given in Table). ECLS support was needed in these patients because of postoperative RV failure. Conduit re-replacements were performed after diagnostic cardiac catheterization due to conduit dysfunction with high RV pressures. On inspection, the conduits were uniformly dilated and thrombosed (one completely, two partially). Degeneration, distal stenosis or dissection was not detected. After the surgery all of them were successfully weaned from ECLS. Postoperative course was unremarkable.
Conclusions: Contegra® conduit is widely applicable to RVOT reconstruction with satisfactory early and mid-term results. However, there is a significant incidence of conduit-related complications, particularly with the younger children (<1 year), smaller conduit sizes (12 and 14 mm) and high pulmonary resistance. The advantage of the Contegra® conduit over a homograft in smaller patients (<1 year) also remains uncertain. High pressure in the conduit may lead to aneurysmal dilatation and eventual valvular regurgitation. However, high pressure in the dilated conduit and valve sinuses may be associated with thrombosis. We would recommend cautious use and close follow-up of the conduits in the early postoperative period in these patients with high pulmonary artery pressures.

P2168 - LATE ARTERIAL SWITCH IN A PATIENT WITH TRANSPOSITION OF GREAT ARTERIES AND INTACT VENTRICULAR SEPTUM

Sertac Haydin[1], Erkut Öztürk[2], Okan Yildiz[1], Behzat Tuzun[1], Alper Guzeltas[2]
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Background: The primary arterial switch operation (ASO) has become the standard treatment of choice for transposition of the great arteries with intact ventricular septum (TGA-IVS) in the first few weeks of life. There is no any standard treatment protocol in patients more than 3 weeks of age.In this report a successful arterial switch procedure with ECMO support was presented in a patient with TGA-IVS.
Case Report: A 30 months old, Syrian male with a body weight of 12 kg was admitted to our clinic. Transthoracic echocardiography demonstrated transposition of great arteries and an atrial septal defect of 10 mm in diameter with colour Doppler. There was a banana-shaped LV involution.The measured LV/RV pressure ratio was 0.5 at the angiography. A decision was taken to proceed with an ASO. Due to insufficient LV contractions and hypotensive progress following weaning from cardiopulmonary bypass (CPB), the patient was switched to ECMO support at the postoperative second hour in the intensive care unit. Neck cannulation was performed. Inotropic support was typically administered as milrinone (0.5 microgram/kg/min) and a low dose of epinephrine (0.05 microgram/kg/min) for the first few postoperative hours. The ECMO support was reduced gradually and the patient was disconnected from the ECMO support at the end of one week.On the following days (postoperative 10th day) the general condition of the patient was good and LV returned to normal shape from the original banana shape. He was discharged without a neurologic sequela on the postoperative 20th day.
Conclusion: Late ASO operation can be performed safely in suitable patients with TGA/IVS after a detailed evaluation and with an efficient, advanced and suitable postoperative intensive cardiac unit monitorization.

P2170 - FATE OF THE LEFT VENTRICULAR OUTFLOW TRACT AFTER RASTELLI TYPE REPAIR IN 107 PATIENTS

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Objective: There are an increasing number of procedures for the correction of aorta arising predominantly from the Right Ventricle with a sub-arterial VSD. Rastelli repair has been subject to longest follow-up but has been criticized for a high incidence of subsequent Left Outflow Tract Obstruction (LVOTO). Resection of the infundibular septum and septomarginal muscle bundles may reduce this risk. This study addresses the risk of early and late LVOTO using Rastelli with muscle resection.
Methods: From 1988 to 2015 all 107 patients undergoing Rastelli-type procedure were studied and divided into, a. Rastelli Procedure in TGA/VSD with pulmonary atresia/stenosis (n = 44), b. Rastelli Procedure in ccTGA/VSD with pulmonary atresia/stenosis i.e. Rastelli-Senning (n = 48), c. DORV Repair in the setting of Pulmonary Stenosis/Atresia with Right Ventricle to Pulmonary Artery Conduit (n = 15).
Results: Median age at index operation was 3.2 years (IQR 1.8-6.3). VSD enlargement was performed in 28 patients. Median follow up time was 8.4 years (Range 0.2–26). 30 day mortality was 3.7% (n = 4) and late phase mortality at 27 years was 10.3%. Pacemaker was required in 7.4% (n = 8). Freedom from LVOT reintervention was 99 (CI 96-100) %, 97 (CI 92-99) % and 90 (CI 88-96) % at 1, 5 and 10 years respectively (Figure 1). There were no early deaths after reintervention for LVOTO (n = 9) and two cases required pacemaker. No patient required a second reintervention. Amongst LVOT reintervention group (n = 9), 6 had not had muscle resected at time of Rastelli.
Conclusions: The incidence of late LVOTO after Rastelli is not high, particularly with attention to muscle resection to enlarge the VSD. Reintervention is safe with no recurrence. The procedure has stood the test of time without destabilizing the aortic root or risking the coronaries. The optimal approach will only be evident when there is longer follow-up of more modern procedures.

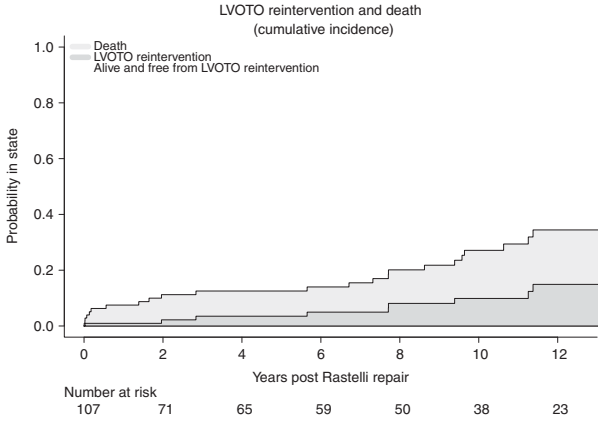

Figure.

P2179 - CONTEMPORARY OUTCOMES OF THE FONTAN OPERATION WHAT HAVE WE LEARNED IN 21 YEARS

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Background: The Fontan procedure is the accepted standard for single-ventricle palliation. Despite improvements in outcomes, ongoing assessment of risk factors for acute and chronic failure is important. We present one of the largest contemporary single-institutional cohorts.
Methods: Patients undergoing a Fontan operation between 1995 and 2016 were included. Failure was defined as death, transplantation, takedown/revision, fenestration creation or enlargement, plastic bronchitis, protein-losing enteropathy, or major perioperative reintervention. Multivariable logistic and Cox regression analyses were used. Data is reported as odds ratios (OR) or hazard ratios (HR) with 95% confidence intervals.
Results: The cohort included 610 patients. Median age at surgery and follow-up time were 4 and 6.8 years, respectively. Diagnoses were varied, with the most common being hypoplastic left heart syndrome (n = 154, 25%), tricuspid atresia (n = 103, 17%), and double-inlet left ventricle (n = 92, 15%). There were 113 patients (19%) with heterotaxy and 50 (8%) with a genetic syndrome or chromosomal abnormality. Surgical trends show increasing use of extracardiac conduits, non-fenestrated Fontan, and early extubation in the operating room. Overall, there were 15 heart transplants and 17 deaths throughout the follow-up period, with three deaths occurring in the perioperative period. Transplant-free survival at 5, 10, and 15 years was 97%, 94%, and 92%, respectively (Figure 1A); freedom-from-failure was 91%, 89%, and 87%, respectively (Figure 1B). Extubation in the operating room was associated with lower risk of acute failure (OR = 0.30, 0.11-0.87). Independent risk factors for chronic failure included genetic syndrome (HR = 2.54, 1.11-5.83), ventricular dysfunction (HR = 3.86, 1.81-8.24), cardiopulmonary bypass (CPB) time in 30-minute intervals (HR = 1.242, 1.100-1.402), and persistent pleural effusions (HR = 4.26, 2.25-8.07). Moderate/severe atrioventricular valve regurgitation (HR = 2.61, 1.13-6.02) and CPB time (HR = 1.223, 1.029-1.452) were associated with reduced long-term transplant-free survival.
Conclusion: Contemporary medium-term outcomes for Fontan patients are reassuring. Lifelong follow-up is mandatory to determine long-term outcomes and need for additional surgery as patients reach adulthood.

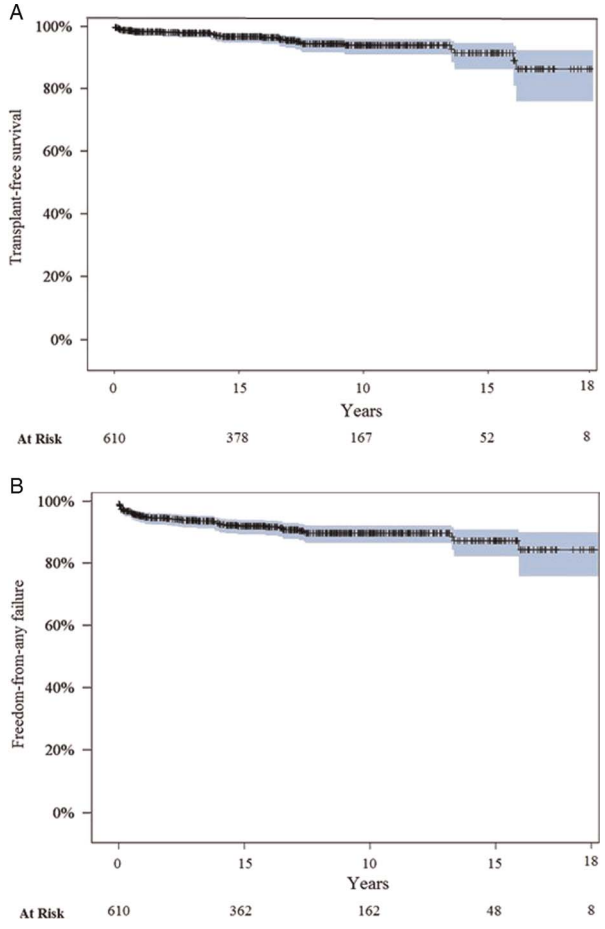

Figure.

P2184 - DURABILITY OF TISSUE ENGINEERED BOVINE PERICARDIUM (CARDIOCEL) WHEN USED FOR THE REPAIR OF CONGENITAL HEART DEFECTS AT 24 MONTHS OR LONGER FOLLOW UP

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Background: To investigate the indications for usage and incidence of failure of CardioCel (Admedus, Fremantle, Australia) when used for the repair of congenital heart defects in the paediatric population.
Material and Methods: Between October 2012 and November 2014, CardioCel was implanted in 130 patients (135 procedures, 194 implants) involving 16 (12%) neonates, 76 (56%) infants and 43 (32%) children >365 days with 99 (51%) simple septation procedures and 95 (49%) more complex three-dimensional reconstructions. All patients have been followed up for 24 months or longer. Implant failure was defined as embolism, infection, thrombosis, structural leak, radiologic evidence of calcification or re-intervention due to patch failure. Six explants underwent histological evaluation.
Results: CardioCel failed in 9 patients (6.7%) (neo-intima/granulation tissue formation in 8; thrombosis in 1). Only 1 implant failed within 30 days (IVC reconstruction; thrombosis). 7 of the 9 failures occurred within 12 months. 8 patients have required 12 re-interventions (6 catheter, 6 surgical). Both implants failed when used for intra-atrial baffle reconstruction in total anomalous pulmonary venous drainage and 2 of 3 implants failed when used for reconstruction of systemic veins. Cumulative freedom from implant failure was 95% (95% CI: 91–98) after 12 months, and 93% (95% CI: 89-97) after 24 months and 36 months. The incidence of failure in neonates was 18.7% (3 of 22 implants), compared to infants (6.6%; 5 of 115 implants) and older children (2.3%; 1 of 57 implants) (p = 0.14). Incidence of failure was higher when CardioCel was used for indications other than simple septation [9/95 (9.4%) versus 0/99, p = 0.001]. Histological evaluation of 6 explants demonstrated evidence of early remodeling.
Conclusion: CardioCel demonstrates good durability when used for the repair of congenital heart defects. The incidence of failure is higher when used for the reconstruction of systemic veins or anomalous pulmonary venous drainage.

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P2185 - LONG TERM FOLLOW UP OF PULMONARY HOMOGRAFT PATCHES USED IN THE AORTIC POSITION IN CORRECTION OF PEDIATRIC CONGENITAL HEART DISEASE

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Background: Pulmonary homograft patches are vital components used in the reconstruction of a variety of congenital heart defects in children, particularly those requiring augmentation of the aorta. Long-term follow-up is important in the assessment of it as a reconstructive material. We present our single-institution follow-up of pulmonary homograft patches used in aortic position.
Materials and Methods: From 2001 to 2013, 70 patients (median age: 6.0 days, interquartile range: 2.0–12.8 days) underwent operative reconstruction for their heart defects which included aortic reconstruction with pulmonary homograft patches as the reconstructive material. Norwood-type aortic reconstruction was performed as the first stage of a single ventricle palliation strategy in 49 patients. Aortic arch hypoplasia (16 patients) with or without VSD and interrupted aortic arch (IAA) (five patients) accounted for the remaining patients.
Results: Complete follow-up (to be completed) was available in 58 patients (82.8%, range 2.9-13.0 years after graft implantation). Thirty-day mortality was 3.4%, and overall ten-year survival was 79%. Orthotopic heart transplantation was performed in four patients (6.9%) and ten-year transplant-free survival was 72%. Catheter-based reintervention was performed in nine patients (15.5%) of whom seven patients (12.1%) had intervention targeted to the area of prior patch reconstruction. Reoperation was performed in 35 patients (60.3%), of whom 21 patients were part of a planned staged palliation scheme. Rates of mortality (0%), reoperation (1.7%), and reintervention (12.1%) attributable to the area of patch intervention were low. No aneurysm formation of the patch areas was encountered.
Conclusions: Our diagnosis-specific survival data is similar with that of other high-volume centers. Our follow-up data shows that pulmonary homograft patches are a robust reconstructive material when used in aortic reconstruction for commonly-encountered pediatric congenital heart surgery diagnoses.

P2186 - THE ANALYSIS OF CONGENITAL HEART SURGERY RESULTS THE COMPARISON OF "RACHS 1 ARISTOTLE BASIC COMPLEXITY SCORE ARISTOTLE COMPREHENSIVE COMPLEXITY SCORE THORACIC SURGEONS AND EUROPEAN ASSOCIATION FOR CARDIOTHORACIC SURGERY MORTALITY CATEGORIES" SYSTEMS

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Background: This study was about the comparison of the surgical results of our clinic according to presumption systems of RACHS-1 (Risk Adjustment in Congenital Heart Surgery), Aristotle Basic Complexity Score (ABC), Aristotle Comprehensive Complexity Score (ACC), Society of Thoracic Surgeons and European Association for Cardiothoracic Surgery mortality categories (STAT MC). Secondly our aim was to compare the affectivity of these systems to predict morbidity and mortality.
Materials and Methods: The classification and risk scoring of sequential 1950 patients who had congenital heart surgery with 4 different systems were performed between 1 October 2012- 31 December 2016. The outcome hospital mortality and morbidity (intensive care unit stay > 7days) was calculated for each category from the four models. The discriminatory ability of the models was determined by calculating the area under the ROC curve and a comparison between the curves of the four models was performed.
Results: The median weight of the cases was 7.2 kg (1.8-80 kg). Among the patients, 53% were male and 47.5% were younger than one year of age. Of 1950 operations, there were 149 (7,6%) deaths, and 541 (27,7%) morbidity (Table 1). For mortality, areas under the ROC curve were 0.803, 0.795, 0.729 and 0.712 for the STS-EACTS MC, ACC, RACHS-1, and ABC scores, respectively. For morbidty, areas under the ROC curve were 0.732, 0.731, 0.730 and 0.685 for the STS-EACTS MC, RACHS-1, ACC, and ABC scores, respectively (Table 2).
Conclusion: STAT MC and ACC systems were successful to predict morbidity and mortality of patients who would have congenital heart surgery and to evaluate the performance of the surgical centers. But STS-EACTS MC was on the fore front due to high feasibility and performance. It is easy, objective and safe for a small number of operations. ABC system had the lowest performance alone. Combinations of systems during valuation will provide the most benefit.

Table 1.

	Mortality*		Morbidity&	
Model/ Method	Areauinder ROC curve	CI 95%	Areauinder ROC curve	CI 95%
RACHS1	0,729	0.689-0.770	0,731	0,706-0,755
ABC	0,712	0.668-0,755	0,685	0,659-0,711
ACC	0,795	0.753-0.837	0,730	0,705-0,755
STS-EACTS MC	0,803	0.768-0.837	0,732	0,707-0,756

Table 2.

Variables	Number n (%)	Observed Mortality	Morbidty n (%)	Predicted Mortality*
RACHS-1				
Not Scored	17 (0,8)	3 (17,6)	3 (17,6)	-
Category 1	220 (11.2)	1 (0,5)	7 (3,1)	0.4
Category 2	838 (42.9)	28 (3.4)	138 (16,4)	3.8
Category 3	578 (29,6)	61 (10.5)	231 (39.9)	8.5
Category 4	246 (12,6)	34 (13.8)	125 (50.8)	19.4
Category 5	-	22 (43.1)	37 (72,5)	-
Category 6	51 (2,6)	149	541	47.7
	1950			
ABC				
Level 1	255 (13)	2 (0.7)	7 (2,7)	<1
Level 2	863 (44)	47 (5,4)	224 (25.9)	1-5
Level 3	538 (28)	43 (7,9)	144 (26.7)	5-10
Level 4	294 (15)	57 (19,7)	166 (56,4)	10-20
	1950			
ACC				
Level 1	231 (11,8)	2 (0.8)	4 (1.7)	<1
Level 2	609 (31.2)	23 (3.7)	121 (19.8)	1-5
Level 3	480 (24,6)	24 (5)	116 (24.1)	5-10
Level 4	630 (32,3)	100 (15,8)	296 (46.9)	10-20
STS-EACTS MC				
1	441 (22.6)	3 (0,6)	32 (7.2)	0,8
2	667 (34.2)	14 (2)	133 (19.9)	2,6
3	333 (17)	29 (8,7)	113 (33.9)	5,0
4	455 (23.3)	73 (16)	221 (48.5)	9,9
5	54 (2.7)	30 (55)	42 (77)	23,1

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P2187 - THE EFFECT OF UPPER BODY CENTRAL VENOUS CATHETER INSERTION ON SUPERIOR VENA CAVA ANATOMY AND SYSTEMIC VENOUS RETURN IN INFANTS WITH SINGLE VENTRICLE

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Background: The upper-body central venous (UBCV) catheter insertion is a widespread technique during pediatric cardiac surgery but might result in obstruction. This complication may especially have serious results in patients with cavopulmonary shunt (CPS). In this study our aim is to evaluate the effect of UBCVC insertion at first operation on superior vena cava (SVC) anatomy and systemic venous return during inter stage period.
Methods: The data of 103 children, who underwent palliation for single-ventricle (Norwood operation = 8, stage 1 hybrid palliation = 8, systemic to pulmonary artery shunt = 29, PA banding = 38, PDA stenting = 20) and consequently pre-CPS cardiac catheterization between 2010- 2016 were gathered retrospectively. Patients were grouped as the ones who had UBCV catheter (group 1) and ones who did not have (group 2). The SVC size was evaluated with SVC index and SVC/Nakata index ratio.
Results: UBCV obstruction occurred in 5 of 34 (14.7%) patients in group 1, and in 2 of 69 (2.9%) patients in group 2 (p = 0.038). There were 6 mortality in group 2 and 3 mortality in group 1 but there was no significant difference in mortality (p = 1.0). The mean duration of catheterization was 9 days for group 1 and this was longer in patients who had UBCV obstruction (11.4 days). The demographic data of the patients are demonstrated in Table 1.
Conclusion: This study showed significantly higher risk prevalence of obstruction in group 1. Although, there was no difference between the two groups in terms of indexes, this result might be due to high risk of venous thrombosis and stenosis associated with upper body central line placement. UBCV catheter insertion in patients with univentricular cardiac anatomy might not be proper. Future studies with larger sample sizes will help to clarify this problem.

Table 1.

	Group l(n = 34)	Group2(n = 69)	p value*
Age, months	13.4 ± 10.5	11,3 ± 3.3	0.26
BSA	0.42 ± 0.1	0.37 ± 0.08	0.024
Weight, (kg)	8.8 ± 3.1	7.4 ± 2.3	0.029
Catheter duration, clays	9 ± 3.01	11.3*3,3	0.019
Bilateral SVCs	7(20.6%)	13(18.8%)	1.0
Venou s obstru ction	5(14.7%)	2(2.9%)	0,038
Right internal jugular	1	1	
Right SVC	1	1	
Left SVC	2		
Left innominate	1		
SVC diameter (mm)	9.4 ± 1.8	8.7 ± 2.2	0.1
Indexed SVC[1]	23.1 ± 5.9	23.8 ± 5.5	0.54
Nakata index(mm/m2)	256.4 ± 120.5	206.4 ± 93.5	0.039
SVC/Nakata index ratio	0.11 ± 0.068	0.14 ± 0.07	0.093

* p value of <0.05 was considered statistically significant
[1]SVC size mm/BSA mm^2

P2189 - OUTCOMES OF THE BLALOCK TAUSSIG SHUNT IN THE CURRENT ERA A SINGLE CENTER EXPERIENCE

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Background: Mortality associated with the Blalock-Taussig shunt (BTS) is high in the current era despite advances in perioperative management. This study was formulated to provide data on [1] current indications, [2] outcomes, and [3] factors affecting mortality and morbidity.
Materials and Methods: A retrospective single center chart review identified 95 children operated between 2004 and 2014. Mortality and major morbidity (cardiac arrest, ECMO, re-operation (any cause), catheter intervention) were analyzed using the Kaplan-Meier method. Risk factor analysis was conducted using Cox's proportional hazard regression.
Results: The median age was 8 (0-126) days, weight 3.1(1.7-5.4) kg. 73% were neonates, another 73% had single ventricle physiology (HLHS excluded), 58% were duct dependent. Pertinent surgical data and outcomes are summarized in the table.At 1-year, survival from a BTS was 82% (95% CI [72.7%, 88.4%]), survival free of major morbidity 61.4% (95% CI [50.7%, 70.5%]). On regression analysis, duct dependency predisposed to mortality (p = 0.01, HR 6.74 [1.54, 29.53]) and composite outcome of mortality and major morbidity (p = 0.04, HR 2.15, CI [1.036, 4.466]) and higher graft index to mortality (p = 0.005, HR 1.07 [1.02, 1.12]).
Conclusions: The commonest indication for a BTS was for children with single ventricle physiology. This is different from other published large databases such as the Society of Thoracic Surgeons' series. This difference could be the result of early complete repair and interventional palliation for children with tetralogy of Fallot physiology. The morbidity and mortality associated with a BTS was considerable, partly explained by a higher risk population. The incidence of pulmonary over circulation was high compared to other published series. Change in technique e.g., sternotomy as default approach was notable, which needs further evaluation. Whether wider adaption of interventional palliation in place of BTS would improve outcomes also requires further investigation.

Table.

Sternotomy	92 (97%)	Hospital mortality	11 (12%)
BT shunt size 3 mm 3.5 mm 4 mm	24 (25%) 66 (70%) 5 (5%)	Interval mortality (from hospital discharge to planned surgery)	6 (6%)
Graft index (shunt cross sectional area (mm^2)/ BSA (m^2)	44.39 ± 8.0 4	Interval mortality (from hospital discharge to planned surgery)	6 (6%)
Shunt size (mm) to body weight (kg) ratio	1.1 ± 0.2	Shunt thrombosis (confirmed on angiogram/surgery)	12(12%)
CPB	11 (12%)	Symptomatic shunt stenosis (from insertion to takedown)	10 (11%)
Pul arterioplasty	17 (18%)	Pulmonary over circulation	28 (30%)
Outcomes		Reoperation (any cause)	12 (12%)
ICU stay (days)	8 (1-268)	Catheter interventions	8 (8%)
Hospital stay (days)	15 (1-294)	Pul arterioplasty at planned surgery	53/73 (73%)

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P2193 - WHAT STIMULATES ENDOCARDIAL FIBROELASTOSIS FORMATION

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Background/Hypothesis: The underlying mechanism of endocardial fibroelastosis (EFE) formation is transformation of endocardial endothelial to mesenchymal cells (EndMT) as we have previously reported but the trigger is unknown. Clinical observation indicates that distension of the left ventricle is directly linked to progression of EFE. We developed an isolated whole heart combined with an endothelial cell culture model to test whether cellular strain induces EndMT.
Materials/Methods: Endothelial cells in culture (n = 3/group) were exposed to 10% uniaxial static stretch for 10 hours (stretch) with or without BMP7 (TGF-β inhibitor) on a gelatin-based drug carrier (drug delivery). Cells treated with media served as controls (control). Immature isolated perfused rat hearts (n = 3/group) were stretched to 30% of their original length for 3 hours (stretch) or treated with BMP7 localized drug delivery into the LV (drug delivery) or perfused only (control). Histological analysis by staining for VE-cadherin or CD-31 (endothelial marker) and αSMA (mesenchymal marker) was performed. Double-staining with both markers was indicative of EndMT. Statistical analysis was performed with ANOVA and Dunn's post hoc analysis. Data are expressed as mean ± SEM.
Results: In isolated endothelial cells in culture and endocardial endothelial cells in a whole heart preparation, stretch induced EndMT, reflected by significantly more double-stained endothelial cells in culture (46 ± 13% of total cell count) and endocardial cells in whole hearts (22 ± 4% of total cell count) compared to controls (cells: 7 ± 2%; heart: 6 ± 2%; $p < 0.05$ compared to stretch). In the BMP7 drug delivery group, significantly less double-stained cells were observed compared to stretch and with similar counts as in controls (cells: 7 ± 2%; heart: 3 ± 1%; $p < 0.001$).
Conclusion: In this study, we could show that mechanical strain induces EndMT. Furthermore, feasibility of localized inhibition of EndMT was demonstrated by BMP7 administration at the site of active EndMT. Successful inhibition of EndMT with BMP7 points toward a TGF-β mediated mechanism.

P2202 - MODIFIED VERSUS CONVENTIONAL ULTRAFILTRATION IN PEDIATRIC CARDIAC SURGERY BEHAVIOR OF THE IONS AND GLUCOSE TIME AND VOLUME TAKEN

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Introduction: The correction of congenital heart diseases whith cardiopulmonary bypass (CPB) always increases capillary permeability and retention of body fluid, leading to edema and dysfunction in brain, heart and lungs. The techniques to reduce the malefic effects of hemodilution are: Convecional Ultrafiltration (UF) during CPB and Modified Ultrafiltration (MUF) performed immediately after CPB.
Objective: The aim of this prospective study was to analyze the behavior of the main electolytes and Glucose in blood and fluid filtered by both techniques and to correlate the time and volume of fluid withdrawn by MUF. Method: Fifty-eight children were operated on, with UF in thirty (G1)and MUF in twenty-eight (G2). K, Na, Mg and Glucose were measured in the blood and the filtered fluid at the end of the procedure.
Results: G1: Glucose and K had similar values in serum and fluid at the end of CPB ($p = 0.887$ and $p = 0.978$). Na presented significant differences ($p < 0.001$), with a lower value of Na in fluid than serum. Mg presented lower values in the fluid($p = 0.001$). UF presented a mean time of 80.26 ± 36.26 and mean volume of 326,66 ± 126.83. G2: Glucose and K presented similar values in blood and fluid ($p = 0.222$ and $p = 0.568$). Lower values of Na and Mg were found in the fluid ($p < 0.001$ and $p = 0.038$). MUF time presented a mean of 14.39 ± 1.87 and the volume of fluid was 193.21 ± 29.07. The correlation between the time of MUF and volume was statistically significant ($p = 0.033$) and positive ($r = +0.405$), suggesting interdependent time and volume.
Conclusion: The results showed that, in UF and MUF, the filtrate has similar values of Glucose and K, and lower of Na and Mg, compared to serum values. MUF can be done choosing either time or fluid volume.

P2214 - MODIFIED IN SITU PERICARDIAL REROUTING TECHNIQUE FOR SCIMITAR SYNDROME REPAIR

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Introduction: Several surgical strategies have been described to redirect right pulmonary venous return to the left atrium (LA) in scimitar syndrome. A major complication of these procedures is pathway obstruction. In 2012, we described an alternative strategy to reduce the incidence of this complication. The in situ pericardium is used as an interposed pathway between the scimitar vein (SV) and the LA. We present a modification of this technique, especially useful in small children with marked malrotation of the heart and preoperative SV obstruction.
Materials and Methods: Patients: Two boys, 10 and 24 years-old, with recurrent pneumonias and pulmonary overflow were submitted to surgery after collateral embolization. The older boy had preoperative SV stenosis. Surgical technique: Through median sternotomy, the right pleural space is opened. After being transected above the diaphragm, the SV is longitudinally opened and sutured side-to-side to a similar incision in the lateral pericardium. Cardiopulmonary bypass with bicaval standard cannulation and mild hypothermia is established. As the LA cannot be reached from the right pulmonary venous pericardial recess, a large posterior right atriotomy is made after aortic crossclamping. Through it, the posterior portion of the atrial septum is detached from its insertion line and sutured to the anterior border of the atriotomy to close the right atrium, leaving below a wide left atriotomy. Finally, the in situ pericardium is sutured to the external surface of the heart around the left atriotomy, creating a pathway between the SV and the LA.
Results: Both patients had fast uneventful recovery, and were discharged 5 days after surgery. Postoperative echocardiogram showed wide unobstructed connection between the SV and the LA.
Conclusion: In situ pericardial rerouting technique combined with atrial septum repositioning can effectively restore normal connection between the pulmonary venous return and the LA in scimitar syndrome, by means of a straightforward, reproducible and low-risk procedure.

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P2236 - DIFFERENT RESPONSES TO PRESSURE VOLUME LOAD IN RIGHT VENTRICLE HYPERTROPHY OF CYANOTIC CONGENITAL HEART DISEASE IN A RAT MODEL

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Objective: Right ventricular (RV) function plays a determinant role in the long-term morbidity and mortality in cyanotic congenital heart disease (CCHD), and it is strictly correlated to residual pulmonary artery stenosis or regurgitation postoperatively. However, the biomolecular changes of myocardial cell under abnormal loading pressure remain unclear. For left ventricular (LV) function, calcineurin-activation is a major factor for the development of pathological hypertrophy. We studied the functional changes of myocardial cell of the chronic hypoxia induced right ventricular hypertrophy in a rat model and the corresponding calcineurin-activation under cellular level.
Methods: Male Sprague-Dawley rats (4wk) were placed in a chamber maintained at 10% O2 for 4 week. The chamber was constantly flushed with room air to maintain low (<0.5%) CO2 concentrations. O2 (10% ± 0.5%) was maintained by a Real-time control system with 100% N2 imported. Then rats were subjected to pulmonary artery banding (PAB), pulmonary regurgitation (PR), and sham surgery (Control). After operations, rats were kept in normal air (21% O2). Six months later, rats were functionally evaluated with cardiac echocardiography (ECG), cardiac magnetic resonance imaging (CMR) and right ventricular pressure measurements. Right ventricular hypertrophy and calcineurin-activation were assessed after sacrifice.
Results: Rats with increased pressure load (PAB) or volume load (PR) of the RV developed similar degrees of hypertrophy, yet revealed different functional and molecular adaptation. By comparison, PAB increased the expression of Modulatory-Calcineurin-Interacting-Protein 1 (MCIP1), indicating calcineurin-activation, and the ratio of beta/alpha-Myosin Heavy Chain (MHC). In addition, PAB induced moderate RV dilatation with normal RV output at rest. In contrast, PR did not increase MCIP1 expression, but increased RV volumes and output obviously.
Conclusions: Pressure and volume load induced different functional and molecular adaptations in RV after chronic hypoxia induced ventricular hypertrophy. These results may give a new direction for the therapeutic strategies of CCHD.

P2246 - SHORT TERM OUTCOME OF SYSTEMIC TO PULMONARY ARTERY SHUNT PROCEDURES IN PAEDIATRIC PATIENTS SINGLE TERTIARY CENTRE CONTEMPORARY EXPERIENCE

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Background/Hypothesis: Systemic-to-pulmonary artery shunts are initial palliative surgical procedures in a wide variety of congenital heart defects with biventricular (BV) or univentricular (UV) physiology. Morbidity and mortality associated with these procedures are not insignificant and they vary widely among institutions and databases reviewed.
Materials and Methods: Between January 2011 and December 2015, 101 patients (53 males, 53%) underwent 125 systemic-to-pulmonary artery shunt operations in a tertiary paediatric cardiac centre. We assessed early post-operative mortality and morbidity and identified risk factors that influence outcome.
Results: Fifty-seven patients (56%) had BV physiology and 44 (44%) had UV physiology. Forty-seven patients (46%) had exclusively shunt-dependent pulmonary blood flow and 54 (54%) had additional pulmonary blood supply. Median age at operation was 64 days (1-2623 days) and median weight 4.3 kg (1.6-18.7 kg). Median size of the shunt was 4mm (3-5 mm) and median post-operative length of stay in the intensive care unit (ICU) was 5 days (range: 1-164 days). There were 8 (7.9%) in-hospital deaths; 1 (1.7%) death in the BV group and 7 (15.9%) deaths in the UV group. There were 2 (3.7%) deaths in the group with additional pulmonary flow and 6 (12.8%) deaths in exclusively shunt-dependent patients. Only 2 (1.9%) patients developed necrotising enterocolitis post-operatively (both in the UV and shunt-dependent group). Spearman's correlation coefficient between age at the time of shunt insertion or weight at the time of shunt and length of stay in ICU was -0.25 and -0.27, respectively.
Conclusions: Short-term mortality following systemic-to-pulmonary artery shunt was markedly lower in patients with biventricular physiology and/or additional pulmonary flow source in our institution. Patient dependent factors other than early shunt patency impact on early postoperative mortality in our series. These findings will serve as a benchmark for currently used transcatheter ductal stenting procedures.

P2257 - THE SANO MODIFICATION OF NORWOOD PROCEDURE REDUCES MORTALITY AT STAGE I BUT INCREASES RE INTERVENTION ON PULMONARY ARTERIES AT STAGE II

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Background/hypothesis: Pulmonary artery (PA) distortion can compromise outcomes of surgical palliation of hypoplastic left heart syndrome (HLHS). We investigated the influence of stage I operative strategy on PA anatomy and outcomes.
Materials and methods: All patients palliated for HLHS (hybrid procedures excluded) between 2005 and 2016 at our institution were included. PA angiography was analyzed by a single reviewer. T-tests for continuous variables and Fisher's exact test for dichotomous variables were used.
Results: Twenty-five patients underwent Stage I with BT shunt (group A) and 15 with a Sano modification (group B), of which 14 to the left, one to the right of the neo-aorta. Median age at surgery was 6 days (range 1-99 days). Mortality up to stage II was 20% and 6.6% in group A and in group B, respectively. Thirty-four proceeded to bidirectional cavopulmonary shunt (BCPS – stage II), 28 having undergone a pre-procedural catheter. Median age at stage II was 114 days (range 30-510 days). Cross-clamp and bypass times were longer in group B, this did not reach statistical significance ($p = 0.2$). Twelve patients (86%) in group B required PA reconstruction compared to 3 (15%) in group A. Pre-stage II angiography revealed a 'Y' shape of the branch PAs in most patients in group B with significant origin stenosis in 9 patients, as opposed to an inverted 'T' shape in most group A with origin stenosis only in one. Mortality at 12 months following BCPS was 15% and 28.5% in group A and in group B, respectively.

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Conclusions: Stage I mortality tended to be lower in group B, but tended to increase at stage II, though the difference did not reach statistical significance at either stage. However, there was a significantly higher requirement for branch PA repair in group B at stage II, perhaps due to Y-shape induced stenosis.

P2258 - MITRAL VALVE REPAIR IN THE FIRST YEAR OF LIFE – OUTCOME AND REOPERATION

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Objectives: Mitral valve repair (MVR) in neonates and infants is challenging and standardized methods of repair are not established. The aim of our study was to review our results of patients undergoing MVR at the age of one year or less.
Methods: Patients who underwent MVR between 1975 and 2016 were identified from our departmental database. Preoperative, surgical, postoperative and follow-up data were analysed with a focus on surgical method and echocardiographic data. End-points of the study were: mortality and mitral valve reoperation.
Results: During the study period, a total of 21 neonates and infants underwent MVR. Mean age was 4.9 ± 2.8 months, mean weight was 5 ± 1.5 kg. Eighteen patients (86%) presented with mitral valve regurgitation and 3 patients (14%) with mitral stenosis. In 17 patients (81%) the etiology of the mitral valve disease was congenital, 4 patients had an acquired mitral valve disease. Surgical repair techniques included closure of a commissure (n = 4, 19%), cleft closure (n = 4, 19%), splitting of papillary muscles (n = 5, 24%) and patch augmentation of a leaflet (n = 2, 9.5%). Two patients died within 30 days (9.5%), no patient died during a median follow-up of 8.3 years [20 days–29 years]. On discharge echocardiography, the size of the left ventricle ($p = 0.008$) and the mitral valve regurgitation had decreased significantly ($p = 0.001$). Seven patients (33%) required a mitral valve reoperation at a median time of 36 days [3 days – 2 years] after initial repair. Reoperation consisted of a re-repair in 3 and a mitral valve replacement in 4 patients. Freedom from mitral valve replacement at 10 years was 78.3 ± 9.8%.
Conclusion: MVR in neonates and infants can successfully reduce the size of the left ventricle and the degree of regurgitation. Overall survival is satisfactory, but reinterventions are necessary in selected patients.

P2267 - SURGICAL IMPACT OF TAILORED STRATEGIES FOR PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM ASSOCIATED WITH DEVELOPED SINUSOIDS

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Background/Objectives: Pulmonary artery atresia (PA/IVS) with developed sinusoidal communication (SC) have a possibility of the RV dependent coronary circulation (RVDCC). Postoperative course with this entity often results in unfavorable course due to coronary ischemia. This study was set to investigate changes of SC through staged surgeries, and assess the efficacy of the tailored managements for this group.
Methods: 11 consecutive PA/IVS patients with SC were enrolled from 2008 to 2015. All patients have developed SC (RVDCC 36%) and hypoplastic right ventricle (RVEDV: 22.5 ± 12.0% of N) and aimed at single ventricle repair. Their average age and body weight at Blalock shunt (BTS) was 45.8 ±15.1 d and 3.1 ±0.3 kg. Severity of SC was graded by SC score at the angiogram before each stage. A week before BTS, lowering preoperative SpO2 (targeted 75-80%) was maintained to avoid heart failure by controlling lipoPGE1 or N2 inhalation while checking BNP level to decide timing of surgery (BNP < 500 pg/ml). At bidirectional Glenn shunt (BDG), Veno-Veno ECMO was used to prevent intraoperative myocardial ischemia derived from decompression of RV pressure. Myocardial ischemia was evaluated at each stage by I-123 BMIPP.
Results: There was no mortality. TCPC were completed in all patients. By the time of BTS, BNP level gradually decreased (max preoperative value: 1562.7 ± 1608.8 pg/ml, at op. value: 503.9 ± 670.7 pg/ml). At TCPC, 3 pts. were operated with V-V ECMO (total score >4), and 8 pts. with V-A bypass. The SC scores are shown in Table. Myocardial ischemia was detected in 5 pts before BDG, but improved in 2 at TCPC.
Conclusions: Tailored managements could contribute to the better results in this study. While regression of SC was shown in some cases, prevention from heart failure at BTS and control of RV pressure at BDG and TCPC are key to improve prognosis.

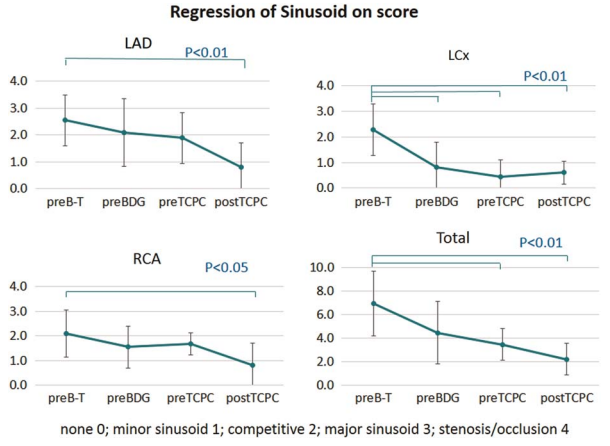

Figure.

P2268 - RISK FACTORS FOR MORTALITY IN REOPERATIONS FOR PEDIATRIC AND CONGENITAL HEART SURGERY IN A DEVELOPING COUNTRY

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Background: The survival of patients with congenital heart disease has increased in the last years, because diagnosis improvement, better surgical techniques and perioperative care. Many patients will require reoperations as part of the stage procedures, grafts deterioration and residual or recurrent lesions. It has been discussed if cardiac adhesions in reoperations consequently increase surgery time, morbidity and operative mortality. The objective of the study was to describe the risk factor for mortality in pediatric patients undergoing a reintervention for congenital heart disease.

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Methods: Historic cohort of patients underwent reoperations after pediatric cardiac surgery from January 2009 to December 2015. Previous surgical approach different to sternotomy were excluded from the analysis.
Results: In seven years, a total of 3.086 surgeries were performed, 481 were reoperations and 238 patients fulfilled the inclusion criteria. Mean reoperation rate was 1.4 ± 0.6 surgeries. Median age at the time of reoperation was 6.4 years. The most common surgical procedure was staged approach for univentricular heart disease (17.6%). Median cross clamp time was 66 minutes. Younger age at the moment of resternotomy, longer cross clamp time and a STAT risk category greater than 3 were risk factors for mortality. The number of re esternotomies was not associated to mortality. No intraoperative mortality was reported. In-hospital mortality was 4.6% and thirty days mortality was 0.54%.
Conclusions: Resternotomy in pediatric cardiac surgery is a safe procedure in our center with low mortality.

P2269 - OPEN CHEST IN COMPLEX CONGENITAL HEART SURGERY WHICH FACTORS AFFECT THE NEW ONSET INFECTION

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Objetive: left the sternum open and packed after pediatric cardiac surgery optimizes systolic and diastolic ventricular function, improve ventilator mechanics and volumes as well as the state of tissue´s perfusion; however, it is associated with increased risk of septic complications. This study was designed to identify risk factors for the development of infection associated with the use of delayed sternal closure in pediatric cardiac surgery.
Methods: Case-control study nested in a cohort of patients with congenital heart disease undergone to surgery. We selected those who were managed with delayed sternal closure. Cases were infected patients while controls were free of it. The association between perioperative variables and the risk of infection was analyzed.
Results: Between January 2010 and September 2016, 3011 patients were operated of which 220 met the study criteria. Median age of 1.4 months (IQR 0.5-5.2). Under one month of age (63.9% vs 36.6%, p = 0.004), stunting (51(48.5-52) vs 54(50-63), p = 0.003) and malnutrition (75% vs 47.2%, p = 0.02) was significantly associated with the development of infection. Risk categories according to disease and type of surgery showed equal distribution between groups. The need for reintervention in the open mediastinum also was strongly associated with infection (30.6 vs 10.6, p = 0.005). Bivariate analysis confirms the association between infectious complications and neonatal age (OR 3.1; CI 95%(1.4-6.6), p = 0.004) and also with the need for preoperative inotropic and/or ventilator support (OR 2.6; CI 95%(1.1-6.3), p = 0.04). On the other hand, a good nutritional state was a protective factor (OR 0.37; CI 95%(0.16-0.86), p = 0.02).
Conclusions: Preoperative factors such neonatal age, low weight and/or height, malnutrition, use of inotropic support and mechanical ventilation along with the need of reinterventions in the open chest represent risk factors for the development of local or systemic infections in patients managed with delayed sternal closure after surgical correction of congenital heart disease.

P2276 - SYSTEMIC TO PULMONARY ARTERY SHUNTING WITH HEPARIN BONDED GRAFTS

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Objective: Systemic to pulmonary artery shunting remains an important palliative procedure for complex congenital heart defects. The use of Heparin-bonded polytetrafluorethylen shunts (HBPS) should decrease the risk of graft occlusion. This study summarizes the results of a single-center after implantation of HBPS in pediatric patients.
Methods: The records of 54 patients treated with HBPS between 2010 and 2016 were retrospectively reviewed. Median age and weight were 14.2 days (range 3-83 days) and 3.25 kg (range 1.8-5.7 kg), respectively. Selected shunt size was 3.5 mm in all except 1 patient (4.0 mm). Seventeen patients (32%) were planned for future biventricular repair and 37 patients (68%) went the univentricular pathway. Shunt modifications included central aorto-pulmonary shunts (n = 35) and modified Blalock-Taussig shunts (n = 19).
Results: Shunt patency until the next surgical treatment was 88 ± 5% after a median duration of 115.4 days (range: 0-241 days). Next surgical treatment in patients with open graft included corrective surgery (n = 9), Glenn procedure (n = 23) and new shunt (n = 5). Patients with subtotal HBPS thrombosis received RVOT-opening (n = 2) and new shunt (n = 4). HBPS take-down due to subtotal graft thrombosis was performed intraoperatively (n = 3), early postoperatively after 3 days (n = 1) and late after 41 days (n = 1). Additionally, 1 patient needed shunt exchange 7 days after Norwood due to thrombosis in the context of failed biventricular repair, and died. Six more patients died (30-day, n = 3) with HBPS in situ, all of them non-shunt related and with univentricular pathology. The 1 and 5 year survival in biventricular patients was 100%, and in univentricular patients 78 ± 8% and 67 ± 8%, respectively.
Conclusions: The use of HBPS is safe and seems to warrant systemic to pulmonary perfusion. Although shunt thrombosis cannot be excluded, patients can be transferred to further palliative treatment, indicating a very slow process of shunt occlusion using HBPS.

P2278 - EARLY RESULTS WITH A STANDARDIZED APPROACH TO RHEUMATIC MITRAL VALVE REPAIR IN PAEDIATRIC POPULATION WITH CUSTOMISED PTFE RING ANNULOPLASTY SINGLE INSTITUTION EXPERIENCE

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Paediatric rheumatic mitral valve disease is still a major health problem in developing countries and mitral valve replacement is associated with problems due to anticoagulation. Our approach now is to aggressively repair the mitral and have been using a fairly standardized approach. For 3 years from January 2013 to 2016 we have done 41 mitral valve repairs with a median age of 11.2 years. Mitral valve is approached through a transeptal modified Dubost approach, posterior leaflet patch is enhanced with pericardial patch (24/41), neochordae to anterior mitral leaflet (24/41) and customised PTFE patch posterior annuloplasty(41/41). There was no

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early or operative mortality but one late mortality on follow up and seven were lost to follow up. There was no hemolysis or thromboembolic episodes in postoperative period. 26/33 is asymptomatic and 6 were in NYHA 2-3 and post op echo showed only 2/33 having more than mild MR. Using a PTFE customised partial ring gives flexibility in choosing the size and extent of the annuloplasty with good early results. Further long term follow up is needed to ascertain how it hold in long term.

P2283 - MITRAL REGURGITATION RISK FACTORS FOR UNFAVOURABLE OUTCOME OF MITRAL VALVE REPAIR TECHNIQUE IN CHILDREN

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Objective: To analyze risk factors for unfavourable outcome (UO) in patients (pts) with mitral regurgitation (MR) that underwent mitral valve repair (MVR) technique.
Methods: Patients with MR who had MVR from 2004 to 2014 were retrospectively evaluated. Mitral stenosis, atrioventricular septal defect and complex cardiac lesions were excluded. UO was defined as reoperation or significant MR (moderate to severe (3+) or severe MR (4+)) during follow-up. Variables were expressed as median. A logistical regression multivariate analysis of predicting factors of UO was used.
Results: Sixty five pts with MR3 + and MR4 + underwent MVR. Etiology was: dysplasia 44.6%, infective endocarditis 13.8%, rheumatic fever 18.4%, abnormal coronary origin 7.7%, others 13.8%. Median age was 8.2 years (ys) (IQR25-75:3.1-13.3) and median weight was 25 kg (IQR:13.5-40.5).Median follow–up time was: 26.5 months (IQR:12-41.5).Ventricular dysfunction was documented in 44.6% and 46.1% had pulmonary hypertension. Surgical procedures were: Annuloplasty (59), cleft closure (19), commissuroplasty (14), leaflet perforation closure (9), leaflet resection (10), chordae replacement or shortening (4), leaflet extension (2), Alfieri (2).Fiftytwo pts (80%) were followed. Fifteen presented UO: 9 were reoperated (7 valve replacement and 2 re-repair). Univariate analysis demonstrated a significant association between UO and: Rheumatic fever (p0.005), preoperative mitral annulus $\geq +5$ SD (p0.002),left ventricle end-systolic diameter $\geq +4$ SD (p0.022), pulmonary hypertension (p0.024) and immediate postoperative residual MR $\geq$ moderate (p0.021). Multivariate analysis demonstrated as independent variables of UO: mitral annulus diameter (p0.012), rheumatic fever (p0.026) and early residual MR (p0.042). No mortality occurred. In mid-term follow-up, Seventy three% remain free of reoperation or significant MR.
Conclusions: A MVR strategy demonstrated a mid-term favourable outcome in children with severe MR, avoiding a valve replacement and its complications. Rheumatic fever, mitral annulus diameter $\geq +5$ SD and immediate postoperative residual MR $\geq 2+$ were predicting factors of UO. Neither age at surgery nor ventricular dysfunction showed statistically significant differences during follow-up.

P2286 - ANATOMICAL REPAIR OF TRANSPOSITION OF GREAT ARTERIES AT HIGH ALTITUDE. A SAFE PROCEDURE WITH LONG TERM SURVIVAL

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Objective: The aim of this study was to present the outcomes of patients with transposition of the great arteries treated at 8,661 feet above sea level.
Methods: Cohort of consecutive patients with transposition of the great arteries treated medically and surgical intervened in a cardiac surgical center in South America located at 8.661 feet above the sea level between February 2003 and November 2016. Descriptive analysis was done by standard statistical tests. A bivariate analysis was performed between patients with or without ventricular septum intact. The center made the follow up by phone and outpatient consultations. Long-term mortality was made for surgical patients. All tests were done in STATA 14.0.
Results: In 13 years, 163 patients with transposition of the great arteries were treated. Secondary to preoperative clinical conditions 20 (12.3%) patients didn't receive surgical intervention and 143 were surgically corrected. Arterial switch was the most frequent procedure 84.6% (121/143). Age at surgery 26 (IQR 8-109 days), Weight at surgery Median 3.4 (IQR 3-4.8 kg), VSD 43.3% (62/143). Preoperative ventilation 21.4%, ICU stay 11(SD 8.2 days). After surgery low cardiac output was the most frequent complication 41%. Open Chest 75.9%, peritoneal dialysis 33.3%. In hospital mortality 8.5%, mortality before 2009, 12.3%, mortality after 2010, 5.6%. At 10 years survival for arterial switch was 93% (95% CI 85-96).
Conclusion: Anatomic repair of transposition of great arteries at high altitude don't affect the mortality and morbidity results and patients older than 3 months have no different outcomes possible due to a altitude related delay in pulmonary vascular resistant drop. Long term survival is good in this topographic conditions.

P2291 - ACUTE HEART FAILURE AND HEMOLYSIS AFTER TRANSCATHETER PULMONARY VALVE IMPLANTATION HEART TEAM APPROACH IN ADULT WITH COMPLEX CONGENITAL HEART DISEASE

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Objective: The aim of this abstract is present the case of a female patient whit congenital valve disease who develop acute heart failure and severe hemolysis after the implant of a percutaneous pulmonary valve.
Methods: 24 years old patient with shone syndrome underwent to Ross procedure in 2001 without complications. In 2010 a percutaneous mitral valvulotomy was performed for severe mitral stenosis. In September 1th 2015 was found a severe stenosis with moderate regurgitation of pulmonary homograft with a NYHA functional class of III-IV, extra institutional transcatheter pulmonary valve implantation was performed. Since post implant period, patient progressed with congestive heart failure and referred to our institution. She also developed intravascular hemolysis, with secondary anemic and jaundiced syndrome, possible related to valve implant.

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Results: Echo showed Ejection fraction 65%, mean pulmonary pressure 80 mm HG, mitral peak gradient 38 mm Hg, mitral mean gradient 14 mm hg, Wilkins index 11/16, aortic valve regurgitation II, membrane sub aortic peak gradient 77 mm Hg. Secondary to severe deterioration with pulmonary edema and mechanical ventilation after 3 days of the admission, a veno-arterial ECMO was implanted. Five days later with hemodynamic stability, percutaneous pulmonary valve was explanted, received a mitral valve replacement with mechanical prosthesis no. 23, right ventricle conduit replacement with by a dacron graft and a bio prosthesis no. 25, tricuspid valve repair, sub aortic membrane resection and ECMO explant, Cross clamp time 134 minutes, pump time 240 minutes. The postoperative evolution was favorable and on November 11th was discharged. On February 2016, secondary to atrial tachycardia and syncope a successful transcatheter ablation was done. Last outpatient clinic visit patient was on October 5th the main findings were NYHA functional class I-II, sinus rhythm, warfarin a beta block treatment.

P2293 - DELAYED TWO STAGE ARTERIAL SWTICH OPERATION. A RENEWED HOPE FOR LATE PRESENTERS WITH TRANSPOSITION OF GREAT ARTERIES AND INTACT VENTRICULAR SEPTUM

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Background: Ductal recanalization by stenting to train the left ventricle has been described for late presenters with Transposition of great vessels, intact ventricular septum (TGA,IVS) and regressed left ventricle. A rapid two stage approach will require a prolonged hospital stay which may drain the resources in developing countries. We report our results of delayed two stage approach in this group of patients.
Materials and Methods: Between February 2010 and April 2016, 16 patients who were late presenters with TGA, IVS underwent successful arterial switch operation through a delayed two stage approach. The median age at ductal stenting was 30 days(13-134 days). The interval between the ductal stenting and arterial switch was 131.75 ± 81.44 days. The median age at arterial switch operation was 136 days (33–671 days).
Results: The median hospital stay following ductal stenting was 5.5 (4.0-7.0) days. The left ventricular posterior wall thickness improved from 0.33 ± 0.04 to 0.41 ± 0.03 mm and the left ventricular mass from 50.44gm/m2 to 60.9gm/m2 after the stenting. There were no hospital deaths after the arterial switch operation. No patient needed post-operative mechanical support. The median hospital stay after switch was 10 (7-22) days. The mean left atrial pressure immediately after surgery was 19.44 ± 2.87 which dropped to 11.44 ± 3.05 before extubation. At a median follow-up of 23(4-76) months, there was one late death. All patients followed up after arterial switch are in NYHA class I and the LV mass increased to 132.21 gm/m2.
Conclusion: Delayed 2 stage arterial switch strategy for late presenters with TGA, IVS helps to train the left ventricle with a shorter hospital stay without need for post-operative mechanical support whereby reducing the financial burden in developing countries. However long term follow-up is needed to assess the left ventricular function.

P2321 - LONGTERM CLINICAL OUTCOMES AFTER FONTAN PALLIATION

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Background: Fontan procedure palliates a diverse range of univentricular hearts. Analysis of the indications, surgical outcomes and follow-up would help us for better clinical outcomes in future.
Methods: Patient demographics, echocardiographic anatomy, hemodynamics, surgical aspects and follow-up were retrospectively studied.
Results: 96 patients aged 10.02 ± 4.61 years (2.6-24 years) underwent Fontan surgery from 2005 to 2016. Common indications being tricuspid atresia (25%), C-TGA (18.75%), Heterotaxy syndromes (14.6%), Single ventricle (13.5%), DORV (12.5%) and d-TGA (10.4%). The mean weight, height and body surface area (BSA) were 26.64 ± 10.5 kg (9-57.5 kg), 132 ± 19.5 cm (75-175 cm) and 0.9 ± 0.26 m2 respectively. Staged Fontan was performed in 88 patients (91.7%) and primary Fontan in 7.3%. Pre-fontan catheterization performed in 93.75% patients revealed following hemodynamic data – mean PA pressure (12.2 ± 2.8 mmHg), LVEDP (11.4 ± 3.4 mmHg), PVRI (2.25 ± 1.04WU.m2). Mean pre-op systemic saturation was 78 ± 6%. 95.8% were extracardiac conduit fontan, 1 was lateral tunnel and two were extra-intracardiacfontan procedures. (16% off pump; 91% fenestrated).Post-operative complications were noted in 45% patients – take down of fontan in one, persistent pleural collection/chylothorax (26%), neurologicalevents (11%), low cardiac output (11%) need for reexploration (9%) sepsis (8%), arrhythmia (7%), diaphragm palsy (3%), gastrointestinal complications (3%), revision surgery for conduit thrombosis/kink (3). Recovery parameters included ICU stay (4.5 ± 3.7d), ventilator duration (17.7 ± 15.3 h), average saturation (90 ± 5%)and CVP (13.9 ± 2.1 mmHg) in first 48 hrs, total ICD duration (8.6 ± 6.7d) and total hospital stay (14.9 ± 8.9d).14% patients required prolonged diuretic therapy (>6 m), while anticoagulants are continued for 85% of followed-up patients (91% of total). Mortality rate was 11.5% with 9 hospital deaths. In mean follow up duration of 4.27 ± 3.12yrs (1m-11.9yrs), 12 required interventions (diagnostic-5, fenestration device closure-3, MAPCA embolization-2, fenestration creation/stenting (2). 83% patients are in NYHA functional class I, rest Class II; 84% continuing studies, 6% dropouts, 3% married, 7% employed.
Conclusion: Proper patient selection, good surgical technique and regular follow up is mandatory to assure good functional long-term outcome after Fontan Palliation.

P2340 - OUTCOME OF IMPROVEMENT STEPS IMPLEMENTED TO REDUCE RISK OF INFECTION DURING CARDIAC HOMOGRAFT PROCUREMENTS

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Background/Hypothesis: Reducing the risk of infections during cardiac homografts procurements is essentials as donors are scarce. Aim was to embark on prospective measurable steps, taken to reduce risk of infections at every stage of cardiac homografts procurement procedure and analyze the outcomes.

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Materials/Methods: A 10 years analysis (2007-2016) procurement of cardiac homograft from fresh cadaveric donors (within 24 hours of death). Procurement performed in the operating theater or mortuary. Donors screened negative cultures for blood and bodily and serology screening of Hepatitis A,B,C, HIV and Syphilis. Cause of death not attributable to infective state. From 2014, improvements steps implemented to reduce infective risk at each stages. Preferences for operating theater settings, strict aseptic harvesting techniques; ensuring patient repainted with povidone iodine and change of surgical drapes after other organs procurements. Utilization of a clean set of surgical instruments and stringent aseptic techniques in triple layering/sealant of organ bags. Homografts trimming in aseptic tissue banking facilities. At each stages, cultures taken from transport solutions, tissues and donors swaps. Penicillin and gentamycin antibiotic into homograft solutions.
Results: A total of 214 cardiac homografts procured (103 pulmonary, 111 aortic homografts). From 2007 to Dec 2013, 26 (12.1%), 13 pulmonary and 13 aortic had positive cultures from solutions and/or tissue. Positive cultures includes Klebsiella 7 (3.3%), Candida 5(2.3%), Acinetobacter 4(1.8%), Streptococcus 2 (0.9%) and others from pseudomonas, VDRL and hepatitis B. From 2014 till 2016, with implementation of new measurable steps, our infection rate is 0%.
Conclusions: Improvements process had successfully reduced procurement infection rates, retaining precious donor hearts, indirectly improves cost efficiency and promoting utilization of cardiac homografts without hesitations or fear of transmission of infections.

P2357 - TEN YEARS SURVIVAL AFTER FONTAN OPERATIONS EXPERIENCE FROM TERTIARY CENTER IN INDONESIA

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Background: Fontan operation is the common procedure performed for uni-ventricular anatomy. Late outcome after Fontan operation is a matter of concern. The aim of study is to evaluate 10 years survival and late morbidity after Fontan operations at National Cardiovascular Center Harapan Kita, Jakarta, Indonesia.
Methods: A retrospective study was conducted from April 2006 to August 2016. All patients surviving to hospital discharge after Fontan operations were included and followed-up until September 2016. Death, and late morbidity were evaluated during follow-up period.
Result: There were 148 patients who underwent Fontan operation during study period. Fourteen patients died post-operative and 23 patients were lost to follow up. Hundred eleven patients with median age 6(3-22) years old were included. Median follow-up period was 895(15-3816) days. Previous palliative operations were carried out in 98(88.3%) patients. Fontan with and without fenestration was performed in 97(87.4%) and 14(12.6%) patients respectively. A total of 8(7.2%) patients died during follow up. The 10 years survival rate was 92.8%. The leading cause of death includes heart failure (50%), infection (25%), arrhythmia (12.5%) and others (12.5%). Late morbidity occurred in 69(62.2%) patients. A total 36(32.4%) patients experienced arrhythmia and 5 (4.5%) required pacemaker implantation. Thromboembolic events occurred in 3(2.7%) patients while 7(6.3%) patients suffered from bleeding (gastrointestinal bleeding (42.9%) and intracranial bleeding (28.6%)). Six (5.4%) patients had right heart failure, 7 (6.3%) with congestive heart failure while 7(6.3%) patients suffered from protein loosing enteropathy. Conduit thrombosis was found in 2(1.8%)patients. Percutaneous ballooning of fontan conduit was performed in 1(0.9%) patient, whilst pulmonary artery stenting was inserted in 1(0.9%) patient. Re-do operation were performed in 4(3.6%) patients. Re-hospitalization rate was 25.2%, with arrhythmia as the common cause.
Conclusion: The rate of 10 years survival after Fontan procedure is satisfactory, although morbidities in the long run are still sub-optimal.

P2366 - TOTAL ANOMALUS PULMONARY VENOUS CONNECTION 25 YEAR EXPERIENCE SINGLE CENTER

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Background: Total anomalous pulmonary venous connection (TAPVC) accounts for 3-4% of congenital heart disease. In the last 20 years there has been a considerable improvement in mortality rates of patients undergoing surgical repair. This study aims to review postoperative results of TAPVC repair over a 25 year period.
Materials and Methods: Retrospective chart review of all cases of TAPVC repairs undergone at a single center from Jan 1991 to Dec 2016. Patient demographics, operative variables, and postoperative course were analyze.
Results: 72 patients with TAPVC were reviewed. One patient died before surgery. Forty-seven (65%) were males. Median age and weight at operation were 2.8 months (range 1 day to 30 months) and 4 kg (range 2.1 to 10.9 kg), respectively. Forty-nine cases (69%) were supra-diaphragmatic and 22 (31%) infra-diaphragmatic. Drainage obstruction was present in 34 patients (47%), 21 (61%) being infra-diaphragmatic. Thirty-one (43%) were operated in the neonatal period, twelve died (38%). Single-ventricle and heterotaxy syndrome, was present in 7 cases (9%) and all of them died after surgery. Post-operative pulmonary venous obstruction occurred in 12 patients (17%), 7 were operated in the neonatal period, 4 had mix drainage and 5 were infra-diaphragmatic. Ten underwent re-intervention and only one survived. Average days of mechanical assisted ventilation were 6.5 and hospital length of stay was 15.6 days. Overall mortality was 31% (35% during the first 15 year period and 20% in the last 10 years).
Conclusions: There has been a significant improvement in mortality rate in the last ten years. Neonatal age, complex congenital heart disease and obstruction previous to surgery, were associated with bad outcome. Post-operative pulmonary venous obstruction had a high mortality rate, and it was associated with neonatal surgery, infra-diaphragmatic and mixed types of drainage.

P2370 - MIDTERM OUTCOMES OF ROSS AND ROSS KONNO PROCEDURES IN INFANTS

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Background: Infants with severe left ventricular outflow tract obstruction or valve regurgitation may require replacement of the

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aortic root with pulmonary autograft. We present our experience ofthe Ross and Ross–Konno procedure in infants.
Methods: There were 8 infants with aortic stenosis operated in our centerbetween January 2015 and September 2016. Three of them underwent aortic commissurotomy and five infants underwent the Ross or Ross-Konno procedure. The median age at the time of Ross or Ross-Konno procedure was 121 days (rangedfrom 38 to 215 days). In the newborn period one patient had aortic commissurotomy and one patient had aortic commissurotomy with the arch reconstruction. They were repeatedlyadmittedto our hospital with aortic restenosis and we performed Ross procedure at the age of 5 and 7 months old, consecutively. One patientof 38 days oldwas operated with aortic commissurotomy. TEE showed severe aortic regurgitation at the operating room. That is why we immediately performed the Ross procedure. An indication for the Ross-Konno procedure was severeleft ventricular outflow tract obstruction in 2 patients.The patients had aortic valve replacement with pulmonary autograft and pulmonary homografts placed in the right ventricular outflow tract in both cases.
Results: Follow-up was available for all the patients (median 189 days, range from 51 to 401 days). There were no deaths during early and late median follow-up. Echocardiography demonstrated good valve function. We have no indications for reinterventions after the Ross or Ross-Konno procedure.
Conclusions: The Ross and Ross-Konno procedurescarry low early and mid-term mortality with low reintervention rate. Long-term results should be evaluated over time.

P2383 - THREE DIMENSIONAL PRINTING IMPROVES MANAGEMENT IN INFANTS WITH COMPLEX CONGENITAL HEART DEFECTS

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Background: Cardiac 3D printing produces highly accurate replicas of a patient's anatomy. Infants with complex congenital heart disease (CHD) represent the highest risk surgical category. We describe the use of 3D printing in infants for surgical planning and patient counseling.
Materials and Methods: Infants (less than 12 months old) underwent cardiac computed tomography (CT) or magnetic resonance imaging (CMRI) for clinical evaluation. CMRI was performed on a 1.5 Tesla scanner with a 3D respiratory navigated FLASH sequence after 0.03 mmol/kg blood-pool gadolinium contrast, gadofosveset trisodium. Contrast-enhanced cardiac CT was performed on a 128 slice dual-source CT scanner. Segmentation, post-processing and 3D printing were performed in collaboration with 3D Systems - Healthcare (Golden, CO) and printed on a ProJet 660 color jet printer. Models for surgical planning were graded for accuracy by two CT surgeons (0-5, 5 = very accurate), who also reported the utility of the models.
Results: 3D models were printed for thirteen infants (age range 1 day – 10 months) with a wide spectrum of congenital heart defects, vascular and related airway abnormalities (Table 1, Figure 1). 8/13 models were used for pre-procedure surgical planning. All pre-surgical models received a score of 5 (highly accurate) compared to OR findings. The surgeons reported the models helped understand complex anatomy, decide between surgical options, decrease cardiopulmonary bypass time, and avoid potential complications. Models for five patients with complex anatomy were used for trainee education and family counseling. All families reported the models aided in their understanding of the child's complex heart disease.
Conclusions: Highly accurate 3D models can be made in patients as early as 1 day of life. These models aid in surgical planning and are useful for family counseling.

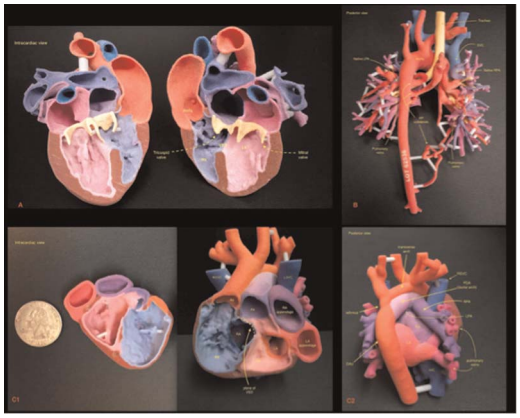

Figure 1.
3D models in infants with complex congenital heart disease. A. Four-month-old (7.7 kg) male with double outlet right ventricle (DORV), pulmonary atresia, large inlet type VSD, bilateral SVCs. The 3D model was printed for pre-surgical planning and shows the relationships between the ventricles, VSD and aorta. B. One-month-old (7.2 kg) male with Tetralogy of Fallot, pulmonary atresia, multiple aortopulmonary collaterals. The pre-surgical 3D model shows two left-sided and one right-sided AP collaterals. These were unifocalized to a RV-PA conduit with model guidance. A separate AP collateral is seen arising from the descending aorta. C. One-day-old (3.1 kg) male with dextrocardia, D-TGA, VSD, long segment arch narrowing, and coarctation of the aorta. The 3D model shows highly precise anatomic detail in a very young infant.

Table 1.

Diagnosis	Age at imaging study (days or months)	Weight at study (kg)	Reason for Printing	Imaging Modality	Accuracy via OR inspection (0-5)
Dextrocardia, D-TGA, VSD, long segment arch narrowing, coarctation	1 days	3.1	Family Counseling Trainee Education	CTA	N/A
Tetralogy of Fallot absent pulmonary valve	4 days	2.3	Family Counseling Trainee Education	CTA	N/A
Double outlet right ventricle, Taussig Bing variant, Interrupted aortic arch	5 days	3.3	Trainee Education	CTA	N/A
Mixed total anomalous pulmonary venous return	16 days	2.3	Family Counseling Trainee Education	CTA	N/A
Tetralogy of Fallot, pulmonary atresia, multiple aortopulmonary collaterals	1	7.2	Surgical Planning Family Counseling Trainee Education	CTA	5
{I,D,X}, Viscero-atrial situs inversus, atrio-ventricular discordance, RV to aorta with pulmonary atresia (double outlet right ventricle). S/p central shunt, alpha-1 antitrypsin deficiency.	3	5.8	Surgical Planning Family Counseling Trainee Education	MRI	5
Supero-inferior ventricles, non criss-cross inflows, large ASD & VSD, LPA stenosis/kinking, arch hypoplasia, left lung collapse	4	5.9	Surgical Planning Family Counseling Trainee Education	MRI	5
Double outlet right ventricle {S,D,D}, non-committed VSD, aorta remote from LV	4	7.7	Surgical Planning Family Counseling Trainee Education	MRI	5
Shone's complex; supra-mitral ring, mitral stenosis, LVOT obstruction, arch hypoplasia. Absent SVC. S/p arch reconstruction followed by resection of supra-mitral ring, MS repair. Persistent MS	5	5.9	Surgical Planning Family Counseling Trainee Education	MRI	5

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Table 1. *Continued*

Diagnosis	Age at imaging study (days or months)	Weight at study (kg)	Reason for Printing	Imaging Modality	Accuracy via OR inspection (0-5)
{l,D,D}, Heterotaxy, viscero-atrial situs inversus, ASD, DORV, D-malposed great arteries, severe PS.	6	7	Family Counseling Trainee Education	CT	N/A
Tetralogy of Fallot, pulmonary atresia, multiple aortopulmonary collaterals	6.00	6.00	Surgical Planning Family Counseling Trainee Education	CT	5
Dextrocardia, viscero-atrial situs inversus, double outlet right ventricle (DORV), L-malposed great arteries, S/P PA band and PDA ligation.	8	8	Surgical Planning Family Counseling Trainee Education	MRI	5
Tetralogy of Fallot, pulmonary atresia, multiple aortopulmonary collaterals	10	6.2	Surgical Planning Family Counseling Trainee Education	CT	5

D-TGA: dextro-transposition of the great arteries, VSD: ventricular septal defect, DORV: double outlet right ventricle, ASD: atrial septal defect, LPA: left pulmonary artery, LVOT: left ventricular outflow tract, SVC: superior vena cava, MS: mitral stenosis, PS: pulmonary stenosis

P2413 - MODELLING DOUBLE OUTLET RIGHT VENTRICLE FOR SURGICAL PLANNING NEW TOOLS IN THE BOX

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Background: Complex forms of DORV pose a challenge for planning optimal surgical approaches. Advances in three dimensional (3D) imaging have enabled the creation of highly realistic computational models of intra- and extra-cardiac structures. In this study, we report the early results of a single centre experience in which patient-specific models were integrated in the decision making process for improving the personalized management of patients with DORV.
Materials and Methods: A cohort of 8 DORV patients diagnosed with non-committed VSD was included in this study. Multimodality images were available for each patient including computed tomography, magnetic resonance and echocardiography. Images were post-processed to: i) recreate virtual 3D models; ii) print 3D replica of the anatomies; iii) perform in vitro surgery; iv) and simulate the virtual haemodynamic conditions following the surgical repair (Figure 1). Where possible, intraoperative surgical morphology was video recorded for post hoc correlation with the virtual model.
Results: All patients had initial palliation in the form of PAB with additional aortic arch repair in 2, PDA ligation in 1 and Blalock-Hanlon septostomy in 1. Six patients underwent a definitive biventricular repair, consisting of creation of a LV-AO tunnel and PA debanding in 4, and arterial switch operation with VSD closure towards the neo-aorta in 2. Two patients were treated along the univentricular pathway. There was no mortality; all biventricular repairs had no LVOT significant gradient at follow-up. Post-procedural peak velocity calculated by computational simulations was found to be in accordance with echocardiographic assessment.
Conclusion: Virtual models allow for good spatial orientation, planning and prediction of surgical approaches in complex DORV surgery. The promising results of this preliminary experience seem to advocate the introduction of this new tool in planning complex CHD surgeries, although further evidence on the benefits of physical models is needed on a larger series of cases.

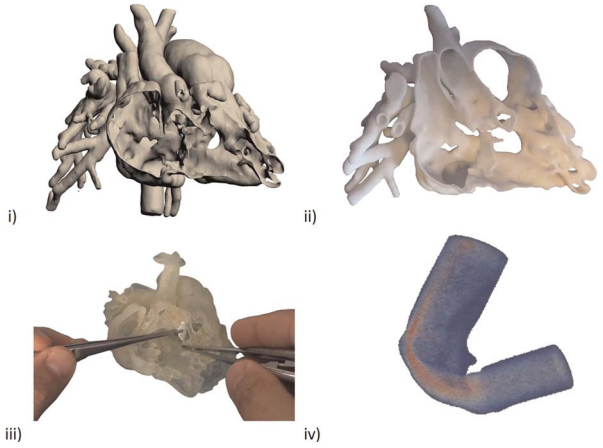

Figure.

P2416 - VIRTUAL 3D SURGICAL TREATMENT OF CONGENITAL HEART DEFECTS

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Background: 3D cardiac modelling has become very popular in pediatric cardiac surgery as it allows a better understanding of complex intra-cardiac morphologies and helps to plan the surgical repair. Despite the additional information given by the 3D models, surgeons are still matching their personal experience with the intra-operative findings to accomplish the surgical repair. We introduced the virtual 3D simulation to either palliate/repair congenital heart defects and then we compared them with the intra-operative findings.
Materials and Methods: Six patients with congenital heart defects who required pre-operative CT scan were included in this study. From the available datasets, 3D segmentation was performed. The models were hollowed and the surgical palliations/repairs were simulated by creating shunts and patches, or by reassembling the intra-thoracic structures. Dimensions and shapes of the patches, and the length of the shunts were collected and used to recreate them in the surgical field.
Results: Seven procedures (in 6 patients) were 3D virtually simulated on the computer and then performed in operative room. Patches were 3D virtually simulated to repair: - common arterial trunk (1 patient) - pulmonary atresia with ventricular septal defect (1 patient) - right partial anomalous pulmonary venous connection (1 patient). Right modified Blalock-Taussig shunt for absence of right pulmonary artery (1 patient) Refashioning of the intrathoracic vessels (2 patients) In all patients who underwent surgical palliation or repair following the 3D virtual computer simulation, the patch and shunt materials, tailored based on the shape provided by the 3D virtual modelling, resulted absolutely precise and didn't require any adjustment. All surgical procedures were uneventful.
Conclusions: The initial experience of this innovative technique of 3D virtual surgical modelling proved to be easily reproducible and reliable for clinical application.

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P2420 - A NOVEL ADJUSTABLE PULMONARY ARTERY BANDING DEVICE TESTED IN 3 KG RABBITS

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Background: Banding of the pulmonary artery is carried out in two circumstances: (i) to prevent excessive pulmonary blood flow in patients with left-to- right shunts, thus preventing pulmonary vascular disease, and (ii) in congenitally corrected transposition of the great arteries where banding is used to retrain the left ventricle and/or treat tricuspid valvar regurgitation. Banding is challenging in determining the optimal degree of constriction such that further surgical interventions may be needed for band adjustment with significant postoperative morbidity and mortality. This challenge has been addressed by the introduction of various banding devices that are adjustable without the need for further sternotomies. These are associated with reduced early morbidity and mortality compared to conventional banding. However, none of these adjustable devices have gained widespread clinical use.
Objectives: To test a novel adjustable device in rabbits.
Materials and Methods: We developed a bidirectionally adjustable pulmonary artery band, using a ball chain that is exteriorised through the ribs and is fixed in a small box placed under the pectoral muscle. Should band adjustment be needed, this may be achieved simply by reaching and opening this box and modifying the length of the ball chain. The band materials permit easy follow-up by echocardiography. This device was tested in 3-kg rabbits during 24 days to see whether or not adjustments were easily feasible under echocardiographic control.
Results: Multiple adjustments were carried out without opening the chest, with simultaneous echocardiographic imaging showing correct device function.
Conclusion: We were able to demonstrate correct device function in an animal model with similar features to those of neonates. Further work is needed to consolidate our findings..

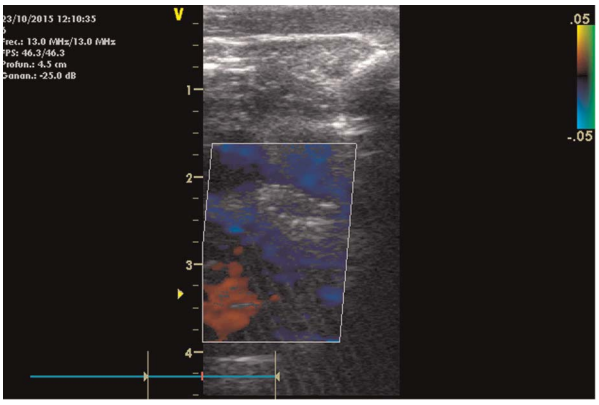

Figure 2.

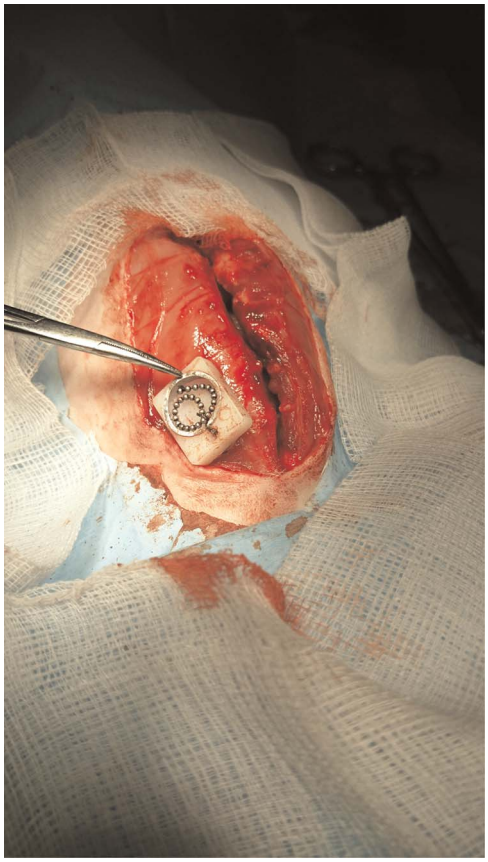

Figure 1.

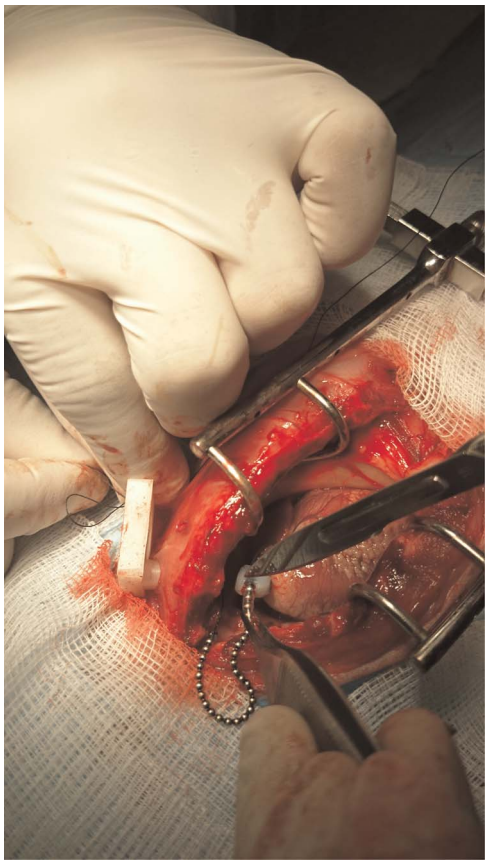

Figure 3.

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Figure 4.

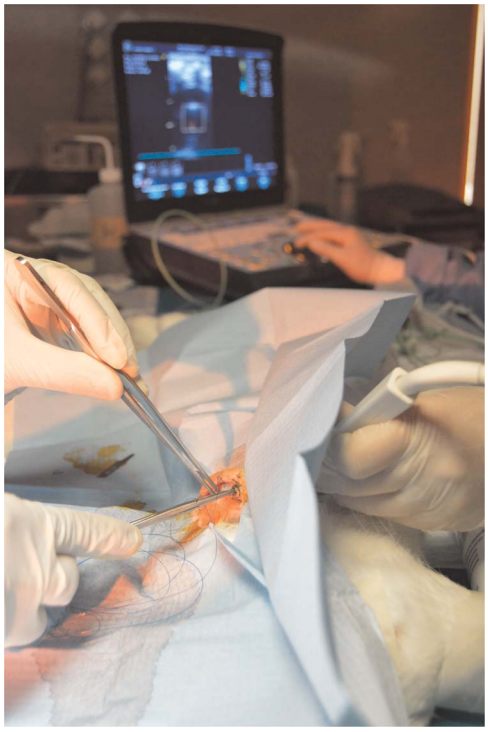

Figure 5.

P2432 - IMMEDIATE RESULTS OF ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY (ALCAPA) CORRECTION

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Background: ALCAPA, also known as the bland-white-garland syndrome, is a rare congenital coronary anomaly correlated with high early infant mortality and sudden death in adulthood. If intercoronary collaterals fail to develop, survival is highly unlikely due to severe myocardial ischemia or congestive heart failure unless corrective surgery takes place.
Material and Methods: From May 2007 to August 2016, four patients diagnosed with ALCAPA were operated at our service. Two female and two male. Weight varied from 5 to 78 kg (m = 48.7), age ranged from 5 to 456 months (m = 235). All patients had echocardiography performed, but it was the coronary cineangiography that lead to the diagnosis in all cases. Three patients also had myocardial scintigraphy and coronary angiotomography, which allows identification of the exact location of the coronary origin. In three of the patients, the left coronary artery (LCA) originated from the right-hand pulmonary sinus. Two of them were submitted to direct reimplantation of the anomalous LCA and the third to the Takeuchi technique. The last patient had the LCA originating from the nonfacing pulmonary sinus. Ligation of the ALCAPA at its origin and coronary artery bypass grafting with the left internal thoracic artery were performed.
Results: None of the patients suffered transoperative infarction or needed any kind of postoperative assisting device. No complications during or after surgery occurred, and the discharge ranged from 8 to 18 days after surgery (m = 10).
Conclusion: To implement the adequate technique for the ALCAPA correction it is crucial to identify from which sinus the coronary artery originates, and that is the role of the coronary angiotomography since it shows the best anatomic correlation.

P2469 - TIMING AND OUTCOME OF CARDIOTHORACIC SURGERY FOR ELLIS VAN CREVELD SHORT RIB THORACIC DYSPLASIATIMING AND OUTCOME OF CARDIOTHORACIC SURGERY FOR ELLIS VAN CREVELD SHORT RIB THORACIC DYSPLASIA

Devyani Chowdhury[1], Katie Williams[2], Millie Young[2], Aaron Chidekel[3], Catherine Preddy[4], Christian Pizarro[3], Kevin Strauss[2]
Cardiology Care for Children, Pediatric Cardiology, Lancaster-United States[1]; Clinic for Special Children, Pediatrics, Strasburg-United States[2]; Ai Dupont Children, Pediatrics, Wilmington-United States[3]; Ai Dupont Children, Pedaitrics, Wilmington-United States[4]

Background: Ellis-van Creveld syndrome (EVC) is a short-rib thoracic dysplasia common among the Amish population (EVC c. IVS13 + 5 G > T) and characterized by acromelic limb shortening, polydactyly, restrictive thorax and an increased incidence of congenital heart disease (CHD), particularly endocardial cushion defect. There is a high postoperative morbidity and mortality following cardiac surgery.

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Methods: Clinical data from 51 EVC homozygotes born between 2005 and 2014 and managed at a single center was reviewed. Postoperative outcome for a subset of 18 children who underwent surgical correction of CHD is presented.
Results: 9 children born between 2005-2009 had corrective cardiac surgery at age 1.3 ± 1 months, whereas surgery was intentionally delayed until 50.1 ± 40 months (P = 0.009) for 9 children born between 2010 and 2014. Compared to the 2005-2009 cohort, delayed surgery was associated with reduced ventilator dependence (1.1 ± 2.4 days vs 49.6 ± 57.1 days, P = 0.075), fewer intensive care days (16 ± 24 days vs 48.6 ± 44, P = 0.155), (fig. 1) and complete prevention of tracheostomy (vs 60%, P = 0.028) and postoperative death (vs 44%, P = 0.082). Relative to age-matched controls subjects, EVC patients had increased respiratory rate that was independent of co-morbid CHD and normalized by 36-40 months of age. (fig. 1).
Conclusion: Delayed surgical repair of CHD in children with EVC decreases postoperative mortality and morbidity. Respiratory rate may be a useful indicator of thoraco-pulmonary maturation.

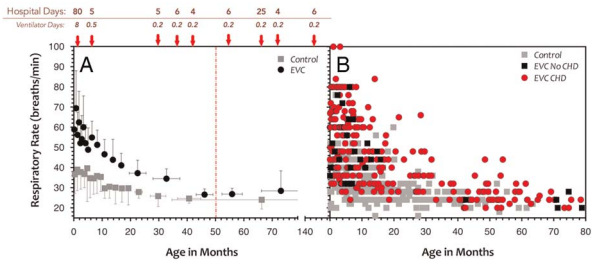

Figure.

P2478 - LONG TERM OUTCOMES FOLLOWING AORTIC ARCH REPAIR IN PAEDIATRIC PATIENTS. A RETROSPECTIVE STUDY

Andrew Ho[1], Khoon-li Kok[1], Genevieve Vanderslott[2], Antonio Ravaglioli[1], Tara Bharucha[1], Nicola Viola[1]
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Introduction: A number of surgical procedures can be adopted to repair coarctation of the aorta, depending on the extent of the lesion. We sought to review medium term reintervention rates and seek risk factors for death or reintervention.
Methods: All patients undergoing procedures from January 2007 to January 2017 involving surgical repair of coarctation of the aorta were retrospectively reviewed. Continuous variables were compared with the Student's t-test and binary variables with Fisher's exact test. Multivariable analyses were performed by logistic regression. A p value of <0.05 was regarded as statistically significant.
Results: 247 patients (96 female) underwent consecutive repair of coarctation. The median weight at primary repair was 3.5 kg (range 1.5–29 kg) and the median age 11 days (range 1 day to 7 years). 20 children were born prematurely and 27 had chromosomal or other genetic abnormalities. Additional structural lesions were present in142, including septation defects in 94 (66%), hypoplastic left heart syndrome (HLHS) in 33 (23%), and Taussig-Bing anomaly in 12 (5%). Surgical repairs included end-to-end or extended end-to-end anastomosis in 133 (54%), forward subclavian flap in 3 (1.2%), reverse subclavian flap in 21 (8.5%) and full arch patch reconstruction in 90 (36.4%). There were 24 deaths, and 26 reinterventions for recoarctation, overall. Excluding patients with HLHS, survival and freedom from reintervention at five years was 92% (95% CI 87-96%) and 89% (95% CI 84-94%), respectively. There was no difference in survival or reintervention by surgical procedure. Multivariable logistic regression identified HLHS (OR 11, p < 0.001), chromosomal abnormality (OR 5, p = 0.01) and presence of other, non-cardiac comorbidities (OR 8, p = 0.002) as risk factors for mortality, but not for arch reintervention.
Conclusions: Early and mid-term outcomes following surgical repair of coarctation of the aorta are excellent, but around 10% of patients require arch reinterventions within 5 years.

P2483 - MID TERM RESULTS OF THE IN SITU PERICARDIAL REROUTING TECHNIQUE FOR SCIMITAR SYNDROME REPAIR

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Background: The main complication associated with surgical techniques in scimitar syndrome repair is pathway obstruction, which accounts for up to 50% of the cases. We describe our experience with the in situ pericardial rerouting technique.
Materials and Methods: Patients: 7 patients with history of recurrent respiratory infections, right cavities dilation and pulmonary overflow were operated on in our institution in the last 3 years. Mean age: 8 years-old (range: 1.9-20 years-old).
Surgical Technique: a) after median sternotomy, the right pleural space is opened; b) dissection of the scimitar vein (SV) c) the pericardium is opened and the right pulmonary artery is cross-clamped; d) the SV is transected above the diaphragm; e) its left lateral aspect is longitudinally opened; f) large incision in the right lateral wall of the pericardium; g) a wide anastomosis is performed between both incisions; h) CPB with aortic and usual bicaval cannulation and moderate hypothermia is established; i) aortic crossclamping; j) a large vertical left atriotomy is performed; k) the pericardium is sutured to the external surface of the heart, leaving the pulmonary venous return connected to the left atrium through a tunnel made of autologous pericardium.
Results: Mortality: 0%. Mean hospital stay: 7,3 days. No major complications. One patient developed transitory massive right atelectasis, which resolved in 72 hs. Mean follow-up: 25 months. Last echocardiogram showed unobstructed pulmonary venous return, with normal low-velocity blood flow throughout the pathway in all patients. Tomographic 3D reconstruction (4 patients) demonstrated wide elliptic connection between the left atrium, the pericardial sac and the SV. Catheterization (3 patients) also showed unobstructed pulmonary venous return.
Conclusions: This technique seems to achieve a wide connection between the pulmonary venous return and the left atrium, with no evidence of pathway obstruction or thrombosis in the mid-term follow-up.

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P2549 - PRELIMINARY EXPERIENCE WITH THE USE OF POLYTETRAFLOUROETHYLENE MEMBRANE TO AUGMENT THE NATIVE PULMONARY VALVE DURING REPAIR OF TETRALOGY OF FALLOT

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Background: During repair of TOF when a trans-annular incision is required, we reconstructed the RVOT using a polytetraflouroethylene (PTFE) membrane to augment the native valve and create a competent valve in the RVOT to avoid pulmonary regurgitation.
Methods: From March 2013 to December 2016, we reconstructed the RVOT using PTFE membrane in 15 patients who required a trans-annular patch in TOF repair. The age ranged from 10 months to 26 years (median 12 years). The weight ranged from 8 kg to 47 kg (median 23.5 kg). In all patients, the trans-annular incision was made across the anterior cusp of the native valve preserving the hinge mechanism leaving the posterior cusp intact. A PTFE 0.1 mm thin membrane was used to augment the divided leaflet. The length of the patch was measured from the annulus to the most proximal extent of the ventriculotomy. A generous width was taken so that the valve would occupy the entire RVOT. The membrane was sutured to the endocardium of the ventriculotomy all around and distally to the divided anterior leaflet. The reconstructed PTFE valve would co-apt with the native posterior leaflet.
Results: There was no mortality. The median follow up was 20 months. All patients were doing well. 2D echocardiography showed good RV function with preserved volumes. There was no significant gradient in the RVOT. Pulmonary valve regurgitation was mild in 12 patients and absent in 3 patients.
Conclusions: PTFE membrane seems to be a good alternative to reconstruct the native pulmonary valve after a trans-annular incision in TOF repair to avoid pulmonary regurgitation. The thin nature of the membrane makes it pliable to use for suturing. Early experience with this material seems encouraging but long-term follow up is required to demonstrate the prevention of pulmonary regurgitation.

Figure 1.

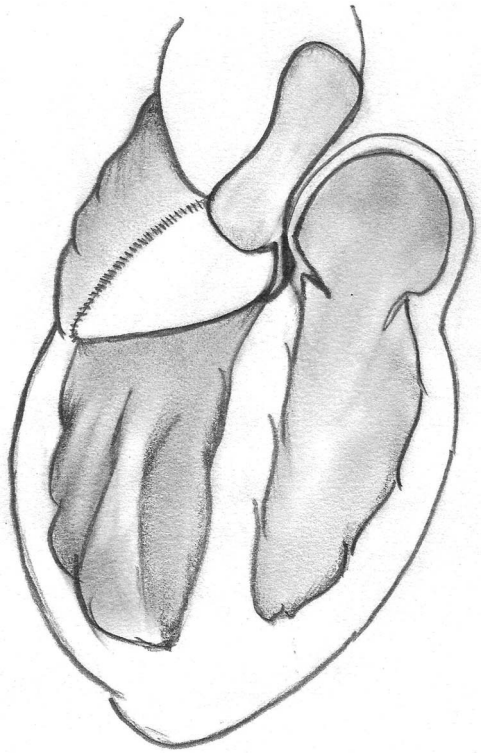

Figure 2.

P2557 - READMISSIONS FOLLOWING CONGENITAL HEART SURGERY AMONG INFANTS AND CHILDREN

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Background/Hypothesis: Thirty-day readmission in after pediatric congenital heart surgery (CHS) has been under studied despite patient vulnerability. We hypothesized that readmissions after pediatric CHS are common and identifiable risk factors exist.
Materials and Methods: We obtained Inpatient Databases for Washington, New York, Florida, and California 2009-2011 and selected CHS admissions age <19 years. We defined readmission as non-elective hospitalization for a patient <31 days of discharge from index CHS admission. We defined complications using the Society of Thoracic Surgeons Congenital Heart complication list. Case mix was adjusted by using Risk Adjustment for Congenital Heart Surgery-1 (RACHS-1). We defined High Resource Use (HRU) admissions as those >90th percentile for total hospital charges. Multivariate analyses using generalized estimating equations estimated adjusted odds ratio (AOR) for readmission.
Results: Among 8,585 index admissions we identified 964 readmissions (11.3%). Median length of stay for readmissions was 5 days, median total charge of $31,973 and mortality rate 1.8%. Among readmissions, 1.6% underwent another CHS of which 44% were HRU, complication rate 88% and mortality 6.25%. On multivariable analysis age 1 month - 1 year AOR 1.3 $p = 0.01$; Hispanic AOR 1.2 $p = 0.03$; government-insurance AOR 1.3 $p = 0.01$; RACHS-1 3 complexity AOR 2.4 $p < 0.001$;

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RACHS-1 4+ complexity 2.0 p = 0.001; HRU AOR 1.4 p = 0.02; complications AOR 1.1 p = 0.04; and emergent index admission AOR 2.0 p < 0.001 were risk factors.
Conclusions: Over 11% of pediatric CHS admissions result in an unplanned readmission. Hispanic race, government insurance, HRU, higher case complexity, complications and emergent index admission are risk factors for readmission.

P2564 - FEASIBILITY OF BIVENTRICULAR REPAIR FOR DOUBLE OUTLET RIGHT VENTRICLE (DORV) NON COMMITTED VENTRICULAR SEPTAL DEFECT (VSD) IN HETEROTAXY AND NON HETEROTAXY SUBSTRATES A CASE SERIES

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Double outlet right ventricle (DORV) physiology and suitability for biventricular repair is based on the routability and location of ventricular septal defect (VSD) in relation to aorta, pulmonary artery anatomy and left ventricle (LV) configuration. We present an analysis of 9 cases with extreme DORV, VSD, decreased pulmonary blood flow physiology , borderline LV and their outcomes after biventricular repair. Over a duration of 2 years, 9 cases of DORV physiology referred for univentricular repair, were committed for biventricular repair based on visual anatomic assessment on table, between the age range of 10 months to 18 years with seven patients doing well on mean one year follow up. Five patients had heterotaxy and four cases had associated complete atrioventricular septal defect (AVSD). In two additional cases, the VSD was perimembranous with inlet extension while the other three had large outlet VSD with perimembranous extension. Five cases underwent right ventricle to pulmonary artery (RV-PA) conduit, and four cases had right ventricle outflow tract augmentation (RVOT) augmentation with transannular patch. Low cardiac output state and prolonged ventilation was observed in 3 survivors, while permanent pacemaker implantation was required for post operative complete heart block (CHB) in one child. None of the cases were reported to have subaortic obstruction. An 18 year old female patient with fontan failure and a diagnosis of transposition of great arteries (TGA) DORV VSD PS, heterotaxy, pulmonary arteriovenous fistulas underwent biventricular repair, but expired due to severe myocardial dysfunction secondary to coronary artery injury. One case with heterotaxy expired 10 months after surgery with severe respiratory sepsis. To conclude Biventricular repair for DORV, non committed VSD is a viable treatment strategy with acceptable early and intermediate term results.

P2576 - OUTCOMES OF SYSTEMIC TO PULMONARY ARTERY SHUNTS IN A LOW INCOME AFRICAN COUNTRY

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Background/Hypothesis: Our unit still uses systemic-to-pulmonary artery shunts for initial palliation of cyanotic congenital heart defects when primary repair is regarded as unsafe. Shunts are thrombogenic and warrant close follow-up; difficult to achieve due to the socio-economic constraints prevalent in a low-income country. Poor follow-up contributes to a delay between the shunt and definitive repair. The study objective is to document the short- to mid-term outcomes.
Material and Methods: Single-centre retrospective review in an established academic unit in a low-income African country. Exclusion: Patients with Hypoplastic Left Heart Syndrome. Primary end points: Mortality (hospital, inter-stage, and at re-intervention); need for re-intervention; definitive repair. Between January 2007 – December 2014,142 consecutive patients underwent a total of 168 MBTS, followed up for 34 ± 27 months (mean ± SD). Age: median 3.1(IQR: 0.9–9.8)months; weight: median 4.5(IQR: 3.3–7.9)kg. Diagnoses: TOF/DORV: 37%; univentricular heart defects (UVH): 18%; pulmonary atresia with VSD(PA/VSD): 15%;TGA: 7%; AVSD with pulmonary stenosis/atresia: 6%; pulmonary atresia with intact ventricular septum (PA/IVS): 4%; miscellaneous: 12%.
Results: Mortality after 1st time shunt procedure (8%), inter-stage mortality (12%) and after 2nd time shunt procedure (5.6%). Cumulative mortality, prior to definitive repair, 20%. Shunt re-intervention prior to definitive repair in 19%. One year survival: PA/IVS 30%, UVH 70%, TOF/ PA-VSD 90%. Definitive correction achieved in only 33% at a median of 18.3 months.
Conclusion: Under conditions prevalent in a low-income country shunts remain a very high-risk procedure with a low yield of definitive repair within a reasonable time frame. Better outcomes may conceivably be achieved through implementing a system of vigilant patient follow-up; structured approaches to earlier definitive repair, or the next step of palliation. The adoption of alternative palliative surgical strategies is justifiable in face of the high overall mortality and warrants exploration.

P2586 - A PROSPECTIVE STUDY TO DETERMINE THE INCIDENCE OF VARIOUS CORONARY ARTERY PATTERNS IN PATIENTS UNDERGOING ARTERIAL SWITCH SURGERY FOR DEXTRO TRANSPOSITION OF GREAT ARTERIES

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Background: Understanding of coronary artery in dextrotransposition of great arteries is necessary for successful outcome of arterial switch operation. The study aims to observe the anatomic diversity of coronary arteries in patients undergoing arterial switch operation, and determine the commonest pattern seen intra operatively in India.
Methods: The study was carried out in a single centre from January 2012 to Jan 2016, 97 consecutive arterial switch surgeries performed during this period were included in the study. We used the Yacoub and Radley- Smith patterns in order to classify the different coronary distributions [A, B, C, D and E] as observed intra operatively.
Results: The commonest coronary pattern seen in the study was type A 57 out of 97[58.76%]. Interestingly, 24.74% of our patients had a type D coronary pattern which is extremely unusual as when compared to what is observed in other parts of the world. This was followed by type B and E [8.24%] each. Type C was not observed in any of the patients.

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Conclusion: In Indian population almost a quarter of the patients present with type D coronary pattern, which is significantly higher than what is observed in other parts of the world.

P2591 - PULMONARY ARTERY STENOSIS AFTER ARTERIAL SWITCH PROCEDURE

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The results of 51 arterial switch procedure were observed. Significant pulmonary artery stenosis in the remote period occurred in 9 patients (17,6%). All 9 patients were examined using CT in addition to routine methods. 6 patients underwent attempts of the balloon plasty of the pulmonary artery stenosis. Only in one case the procedure was successful. We came to conclusions of possible causes leading to the pulmonary artery stenosis: - inadequate dissection of branches of the pulmonary artery results in their low mobility, tension and deformation; - using a xenopericardial patch in the reconstruction of the pulmonary artery; - using of an autologous pericardium treated with glutaraldehyde; - using the trap-door coronary transfer forming increased aortic root which results in the compression of the pulmonary artery and its bifurcation. 8 patients with pulmonary artery stenosis were reoperated using cardiopulmonary bypass. In two patients the stenosis were located at the level of the pulmonary artery anastomosis. In six patients the stenosis were located at the level of the the pulmonary artery branches.
The postoperative period were uneventful. All the patients were disharged with a good results.

P2592 - SURGICAL OUTCOMES AFTER TWO YEARS OF ESTABLISHING A CONGENITAL HEART DISEASE PROGRAM IN A RESOURCE LIMITED COUNTRY USING A 'SINGLE CHARITY CENTRE' APPROACH

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Background/Hypothesis: Guyana, a developing nation in South America, has a neonatal mortality rate of 22.8 and an under five mortality of 39.4 respectively. With no established congenital heart disease or critical care programs, the morbidity and mortality of this group of disease contributes to the country's health burden. The International Children's Heart Foundation (ICHF), a charitable organisation with 22 years of providing paediatric cardiac services in developing countries, undertook the challenge of establishing a congenital heart disease program in Guyana.
Aim: The aim of this study was to evaluate the surgical outcomes over the first two years of introducing a pediatric cardiac surgical program in Guyana and to identify factors transferable to other resource limited countries.
Materials and Methods: This is a retrospective case series review of patients who underwent cardiac surgery by the ICHF team from January 2015 to December 2016.
Results: Ninety-five patients underwent corrective or palliative cardiac surgery. Ninety percent (n = 85) were children, 55% percent (n = 47) were less than 5 years old. Seventy percent (n = 80) had a RACHS-1 score of 2 or more. The range of cardiac diagnoses and interventions were comparable to established programs within developed countries (Table 1). Seventy percent (n = 55) of the cases returned intubated after surgery. The median time of mechanical ventilation on the PICU was 120 minutes (IQR 0, 247.5). Only twenty percent of the patients (n = 18) received analgesic infusions. The most frequent post-operative complication was bleeding (Table 2). In this series, 30 days mortality was 2.1%.The median PICU length of stay (LOS) was 2 days without and 3 days with any inotropic requirements.
Conclusion: Pediatric cardiac surgery can be performed with good outcomes in developing countries. Optimal use of analgesia, shorter periods of invasive ventilation, early mobilization, using an experienced team to supervise and train the local health care professionals may contribute to shorter PICU LOS.

Table 1.

DEMOGRAPHICS		INTERVENTIONS	
Age -yrs (median, IQR)	5.05 (1.7-13.9)	**Surgery:**	**n = 85**
Age below 5 years	47	Single Lesions	41
Weight- kgs (median, IQR)	15.6 (8.5-31)	Complete Tetralogy 0(Fallot Repair	13
Male	42	Single Ventricle Palliative Procedure	9
Malnourised	25	Other complex procedures	22
RACHS-1	20		
RACHS-2	36	CPDTime (minutes) (median, IQR)	100 (72.3-127)
RACHS-3	20	ACX Time (minutes) (median, IQR)	59 (33-86.5)
RACHS-4	4		
Diagnosis	**n = 85**		
Single lesions(ASD/ VSD/PDA/AVSD)	41		
Single ventricle Lesions	11		
Tetralogy of Fallot	13		
Other Complex Cardiac Lesions	20		

Table 2.

Post Operative Care (On Admission) n = 85		complications (n = 37)	
Epinephrine infusion	26	Reintubation	5
Milrinone infusion	50	Reoperation	5
Dopamine infusion	10	Pulmonary Hypertension	5
Analgesia infusion	18	Bleeding requiring transfusion	11
invasive ventilation	55	Temp Epicardial pacing	5
Non inotropic infusion	22	junctional ectopic achy	1
		Delayed chest closure	2
		Surgical site infection	1
		Death	2

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P2603 - AN UNNOTICED PARTIAL PULMONARY VENOUS RETURN COMPLICATING SURGICAL CLOSURE OF A SWISS CHEESE VSD

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PAPVC and VSD co-existence, both causing significant left to right shunt, is exceptionally rarely reported. We herein report a case of Swiss cheese VSD closed surgically with a peroperatively diagnosed PAPVC. To our knowledge there is only one report depicting such a case in the literature.
Material and Method: A 2-year old male with a history of pulmonary artery banding (PAB) in infancy, due to a swiss cheese VSD at another clinic, underwent patch closure of the VSD and debanding at 1 year old. However patient could not be weaned from cardiopulmonary bypass and surgical team made a decision to takedown the VSD patch with a loose re-PAB, but ECMO support to provide adequate systemic organ perfusion was mandatory. Patient was discharged on the 1st postoperative month under pulmonary anti-hypertensive therapy.
Results: Patient was referred to our clinic and transthoracic echocardiography (TTE) revealed a swiss cheese VSD and an uneffective PAB. Cardiac catheterization reported a defect unamenable for catheter closure thus the patient was scheduled for a total correction. Right upper pulmonary veins draining to superior vena cava was noticed peroperatively, therefore correction of the PAPVC and VSD closure was performed (Figure 1). Patient was extubated on the 2nd postoperative day and control TTE depicted no residual VSD with unrestrictive pulmonary vein flow. The post-operative course was uneventful and the child was discharged on the 7th post-operative day in good condition.
Conclusion: Although preoperative diagnostic studies are sine qua non for the congenital heart surgeon, meticulous peroperative evaluation of the patient with his/her unique history is vital for splendid results, exceptionally in redo cases.

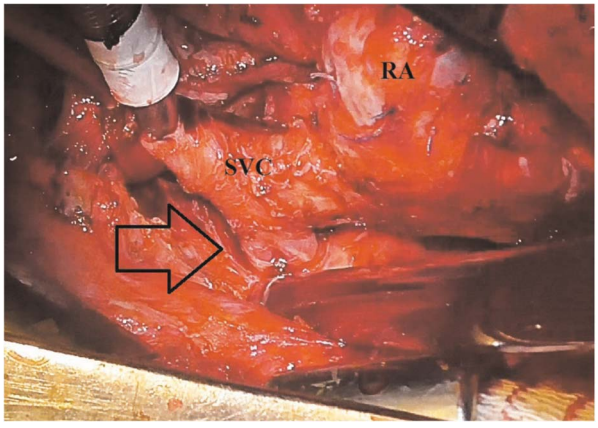

Figure.
Intraoperative photograph showing the cannulated SVC and right upper pulmonary veins draining to superior vena cava (arrow). Abbreviations: SVC, superior vena cava; RA, right atrium.

P2613 - NIKAIDOH PROCEDURE INITIAL EXPERIENCE FROM SINGLE CENTRE

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Madras Medical Mission Hospital, Paediatric Intensive Care, Chennai-India[1]; Madras Medical Mission Hospital, Paediatric Cardiac Surgery, Chennai-India[2]

Background: The Rastelli operation is the conventional procedure for TGA with VSD, PS. Its results, however, are suboptimal. Although short term results are good it has long term complications due to conduit obstruction, arrhythmia. Another surgical technique, the Nikaidoh procedure, appears as a good therapeutic option.
Objective: Review our initial experience using the modified Nikaidoh procedure consisting of aortic translocation and biventricular outflow tract reconstruction in the short term.
Methods: The study analyzed 5 consecutive patients operated on with the modified Nikaidoh procedure at our institution.
Results: The mean hospital stay duration was 15 days (11- 23 days). Ventilatory dependency was noted for mean of 5.4 days (2-9 days) and ionotropy for mean of 7 days (5-11 days). Only one patient developed moderate LV dysfunction. None of the patient had aortic valve regurgitation. One patient developed post operative JET which recovered in three days.
Conclusions: The Nikaidoh operation and its variants are a satisfactory surgical option for patients presenting with TGA,VSD, PS morphology who are inadequate candidates for a Rastelli procedure. In the short term follow-up, the left ventricular outflow tract remained without obstruction and there was no significant aortic valve regurgitation.

P2631 - ON TABLE EXTUBATION FACILITATES MANAGEMENT OF POSTOPERATIVE PULMONARY ARTERIAL HYPERTENSION A NOVEL APPROACH

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Background: Past two decades have seen rapid improvement in outcome for congenital heart surgery after palliation and complete repair. Post operative pulmonary hypertensive crisis is a potentially fatal complication occurring unpredictably in the post operative period in patients with severe pulmonary arterial hypertension. The aim of this study was to evaluate the safety of extubation in the operating room after pediatric cardiac surgery in patients with severe pulmonary arterial hypertension using our institutional protocol.
Methods: A retrospective analysis of 122 patients aged 15 days – 7 years (median 12 months) with severe pulmonary arterial hypertension due to large left to right shunts having undergone corrective surgery was conducted. Patients on preoperative inotropes, emergency surgery, preoperative ventilation and spinal deformity or deranged coagulation profile precluding central neuraxial blockade were excluded from the analysis. Statistical analysis of factors influencing inability to extubate and risk factors were assessed. Induction of anaesthesia with a combination of Ketamine and midazolam, maintenance with sevoflurane, use of vasopressin, epidural analgesia with bupivacaine and postoperative sedation with combination of dexmedetomidine and epidural bupivacaine was the standard protocol adopted. Post extubation high flow nasal oxygen therapy was used in all cases.
Results: In our analysis, 107 out of 122(87%) patients having undergone corrective surgery with lesions with severe pulmonary arterial hypertension were extubated in the operating room. 3

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patients (2.8%) required reintubation due to various reasons but not due to pulmonary hypertensive crisis. There was no mortality in the extubated group. Lower weight, lower age, high inotropic score with unstable hemodynamics and complex repairs with longer bypass time were factors preventing extubation.
Conclusion: On table extubation is a safe strategy in majority of patients after corrective surgery in lesions with severe pulmonary arterial hypertension and reduces the risk of pulmonary hypertensive crisis.

P2635 - UNIFOCALIZATION OF THE MAJOR AORTOPULMONARY COLLATERAL ARTERIES WITHOUT DIRECT ANASTOMOSIS IN PATIENTS WITH PULMONARY ATRESIA AND VENTRICULAR SEPTAL DEFECT USEFUL APPROACH

Joaquin Perez-Andreu[1], Joaquin Fernandez-Doblas[1], Dimpna C Albert[2], Ferran Gran[2], Gemma Giralt[2], Anna Sabate-Rotes[2], Pedro Betrian[2], Queralt Ferrer[2], Ferran Roses[2], Raul Abella[1]
Vall D´hebron University Hospital, Congenital Heart Surgery, Barcelona-Spain[1]; Vall D´hebron University Hospital, Pediatric Cardiology, Barcelona-Spain[2]

Background: The ultimate goal of the pulmonary atresia (PA) with ventricular septal defect (VSD) and major aortopulmonary collateral arteries (MAPCAs) is to create unobstructed and separate in series pulmonary and systemic circuits. The number of lung segments recruited and the status of the pulmonary microvasculature are the main influence factors for the post-repair peak right ventricular (RV) pressure. We present our experience with a 1-stage unifocalization technique that eliminates the need for anastomosis of individual MAPCAs.
Materials and Methods: Between 2008 and 2016,25 patients underwent a 1-stage unifocalization surgery consisting in 1) exclusion of the descending thoracic aorta segment containing the origin of all MAPCAs and 2) connection of this aortic segment to the native pulmonary artery with an interposition polytetrafluoroethylene conduit. The RV was connected to unifocalized pulmonary arteries using a valved conduit. 95% of MAPCAs were finally unifocalized. The median age at surgery was 8 months (range: 4-18). 11 patients needed a rehabilitative procedure of the hypoplastic pulmonary arteries before the definitive repair.
Results: All patients survived the operation with the exception of 1 death in a child with a complete absent of intrapericardial branch

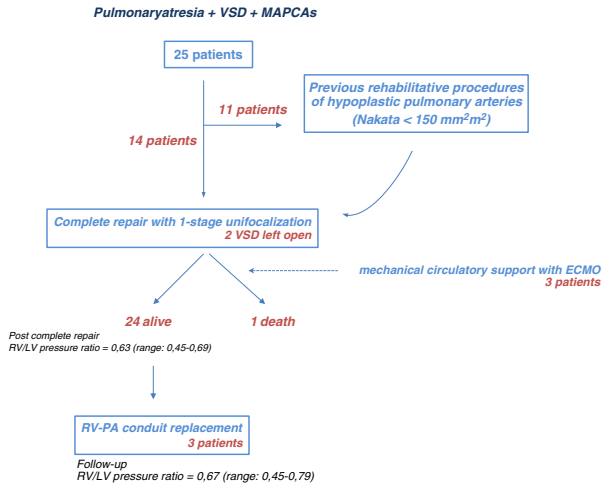

Figure 1.

pulmonary arteries. Postoperative complications included: infection (n = 3), bleeding (n = 2), pleural effusion (n = 1) and diaphragm paralysis (n = 2). The postoperative length of stay was 10 days (range: 7-16). Technetium99m lung perfusion scans in 15 patients one year after the operation showed uniform perfusion of all pulmonary segments. After 8 years of follow-up, all 24 patients are alive.
Conclusions: Complete repair of PA + VSD with unifocalization of all MAPCAs can be successfully accomplished. This approach provides optimal results in selected patients with MAPCAs originating from the descending thoracic aorta..

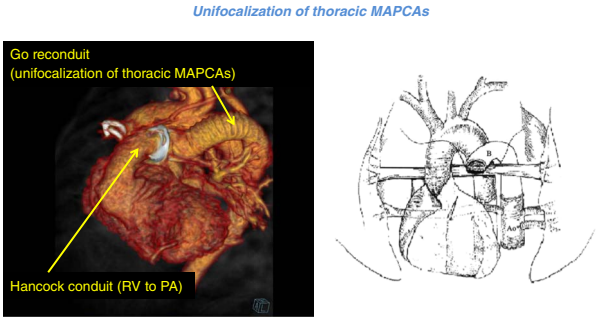

Figure 2.

P2647 - A 6 YEAR SINGLE CENTRE EXPERIENCE ON THE USE OF EXTRACORPOREAL MEMBRANE OXYGENATION IN PAEDIATRIC MYOCARDITIS PATIENTS

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Background: Paediatric myocarditis, although uncommon, carries significant morbidity and mortality if undiagnosed promptly. Extracorporeal membrane oxygenation (ECMO) has been employed as a rescue therapy since 2010 at our institution. We aim to analyze our experience, in terms of the outcome of its use on paediatric myocarditis patients.
Materials and Methods: This is a single centre study in a tertiary hospital on paediatric myocarditis patients treated medically and with ECMO over a 6-year period from Jan 2010 to Oct 2016. Patients were identified from the paediatric intensive care unit (PICU) admission book and institutional ECMO registry. Demographic data was collected retrospectively from the hospital database. The outcome for patient was classified as either survival to hospital discharge or death. Data on complications and ejection fraction (EF) were also collected.
Results: 9 children (3 boys and 6 girls, mean age 9.78 years) were diagnosed to have myocarditis. Two (22%) were medically managed in PICU, and they received both dobutamine and milrinone, and did not require invasive ventilation. They were the two eldest in the study group (ages 15.92 and 17.36 years), and also had a higher EF pre treatment (59% and 72%) and post treatment (73% and 72%). We used veno-arterial ECMO on seven (78%) children, one of whom (girl, age 6.83 years) died on day 15 of admission due to systemic bleeding and severe cerebral oedema. Complications in other patients included haemorrhagic stroke, vocal cord paresis, lung barotrauma, pleural effusion, pneumothorax, urinary tract infection, acute urinary retention, transaminitis, ileus, line

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infection and sepsis. The majority of our myocarditis patients required ECMO, and they did well with a survival rate of 86%.
Conclusions: ECMO has good outcomes in myocarditis. ECMO should be considered as one of the treatment modalities in paediatric patients with myocarditis.

P2659 - LONG TERM OUTCOMES OF JATENE PROCEDURE

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Background: Transposition of the Great Arteries (TGA) is the most prevalent cyanogenic congenital heart disease in the neonatal period. It presents totally unfavorable evolution if it is not corrected surgically, and the Jatene Procedure is the gold standard for the treatment. The aim of this study is to describe clinical and functional characteristics of patients undergoing Jatene procedure during long-term outpatient follow-up.
Material and Methods: Retrospective study of patients submitted to Jatene procedure, in outpatient follow-up at a tertiary service. The following variables were evaluated: type of TGA, performance of Lecompte maneuver, development of arrhythmias during follow-up, need for pacemaker, presence of heart failure, major artery insufficiency or stenosis and residual ventricular septal defect (VSD).
Results: A total of 113 patients were included in the study, 73 (64.6%) were males, with a mean follow-up time of 9.8 ± 8.7 years. The mean age at surgery was 2.8 ± 8.4 months. Regarding the type of TGA, 63.5% were simple. Of the patients in whom the Lecompte maneuver was performed ($n = 81$), 22% evolved with neopulmonary stenosis compared to 47.8% of those who did not undergo the maneuver ($p = 0.033$). There was a need for definitive pacemaker implantation in 8.3% of the patients, signs of HF in 10.7%. Neoaorta dilatation was also observed in 49.1%, neopulmonary stenosis in 30% and neoaorta in 10.1% and residual VSD in 8.1% of patients.
Conclusion: In the long-term follow-up after Jatene procedure for anatomical correction of TGA, the incidence of neopulmonary stenosis was lower in those patients who underwent Lecompte maneuver. Approximately half of the patients evolved with dilation of the neoaorta and a small portion with definitive pacemaker implantation, residual IVC or heart failure.

P2672 - IMPACT OF CARDIAC SURGERY ON MALNOURISHED AANEMIC ANDHHYPOPROTEINEMIC INFANTS TO ADULTS OF INDIA

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Objectives: To study the short and long term effects of cardiac surgery on malnourished, anaemic and hypoproteinemic infants to adults.
Background: There have been several studies but only a few on anemic, malnourished and hypoproteinemic infants to adults.
Methods: This is a Retrospective study of 280 infants to adults with Congenital and Acquired Cardiac abnormalities who had undergone cardiac surgery. There were 212 in the Paediatric age group and 68 in the adult age group. In the pediatric age group, Repair of Acyanotics was in 175, Coarctation of Aorta in 3, Cyanotics in 20, Anomalous Pulmonary Venous Drainage in 4, and Complex Anomalies in 6. Mitral valve Replacement was in 3 and AVR in 1. Among adults 7 Acyanotics had Repair, 42 had Mitral valve Replacement, 3 had Mitral Valve repair, 5 had Aortic valve Replacement, 4 had Double Valve Replacement and 7 had CABG. They have been followed up periodically after 1 month, 3, 6, and 12 months and every year after that with a view to estimate to what extent the preoperative risk factors impact the recovery of vital parameters. Role of each person in family and society is also being ascertained. Follow up has been from 1 month to 12 years.
Conclusion: Irrespective of the fact that they return to the same environment from which they hailed, symptomatically and in terms of growth, they have shown remarkable progress. Return of the parameters however varies. Only long term follow up can reveal the true outcome.

P2673 - REPAIR OF ANOMALOUS RIGHT CORONARY ARTERY FROM THE PULMONARY ARTERY TECHNICAL PEARLS AND PITFALLS

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Background/Hypothesis: Anomalous origin of the right coronary artery from the pulmonary artery(ARCAPA) is rare. Late thrombosis due to technical imperfection has been reported. Unique anatomical characteristics observed are tethering secondary to the extensive large collateral vessels, severe native coronary tortuosity and the side-by-side great vessel orientation which require specific technical considerations to ensure safe translocation.
Materials and Methods: A single center retrospective review of 5 patients with ARCAPA was performed. There were 4 children and 1 adult. Echocardiographic and computerized tomography (CT) scan data were analyzed for anatomical and functional cardiac characteristics. Operative techniques were analyzed which reflected an evolution towards a modified-trapdoor technique.
Results: Patient demographics are listed in table 1. Coronary artery echo characteristics are listed in table 2. CT angiography was used to assess coronary anatomy in all patients and detected a severe ostial stenosis in one. 4 children presented with asymptomatic murmurs and the adult patient with unstable angina. All patients underwent successful surgical correction without complications. The modified trapdoor technique provided the most ideal geometry for coronary transfer secondary to its unique anatomical characteristics. The average hospital stay was 5 days and there were no mortalities. Two patients had coronary button transfers above the sino-tubular junction (STJ) using a vertical stab incisions, one had the button implanted after excising part of the aortic wall and 2 patients had modified trapdoor incisions.

Table 1.

Patient	Age (years)	CPB (minutes)	XC (minutes)	Sex M = male, F = female	Presentation	Translocation Technique
1	4.2	140	73	M	Asymptomatic murmur	Coronary button with Aortic wall excision
2	41.5	208	154	F	Unstable Angina	Patch Augmented button
3	1.8	91	41	F	Asymptomatic murmur	Vertical Slit
4	3.3	188	100	F	Asymptomatic murmur	Modified Trapdoor
5	8.0	155	78	M	Asymptomatic murmur	Modified Trapdoor
Mean	11.8116.8	156 ± 40.5	89 ± 37.5	-	-	-

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Conclusion: ARCAPA is often asymptomatic but can present with ischemia. Its unique anatomical characteristics require a coronary transfer technique different from that performed for aortic root replacement or arterial switch operations. The use of a modified trapdoor incision simplifies coronary transfer and may minimize late coronary thrombosis.

Table 2.

Patient	RCA Z-Score	LMCA Z-Score	LAD Z-Score	Ventricular function	Collateral Flow	Coronary Stenosis
1	1.45	8.4	5.4	Normal	Present	None
2	0.5	3.9	-	Normal	Present	Proximal RCA 90%
3	-	-	-	Normal	Present	None
4	2.7	3.7	2.4	Normal	Present	None
5	5.5	6.3	4.5	Normal	Present	None
Mean	2.5 ± 1.9	5.6 ± 1.9	4.1 ± 1.3	-	-	-

P2674 - NEUROSURGICAL INTERVENTION AFTER CARDIAC SURGERY FOR PEDIATRIC AND CONGENITAL CARDIAC CARE A SINGLE CENTER 17 YEAR EXPERIENCE

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Background: Neurological injury after cardiac surgery can present as seizures, stroke, or hemorrhage. Very few strokes or hemorrhages result in the need for neurosurgical intervention. We reviewed all patients undergoing cardiac surgery at a single institution during a 17 year time interval and identified all patients who required neurosurgical intervention.
Materials and Methods: All pediatric and congenital cardiac operations were identified at a single institution between January 1, 2000 through December 31, 2016 using the institutional software Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database. The records of patients undergoing neurosurgical intervention were reviewed.
Results: 5033 pediatric and congenital cardiac operations were performed. Only two of these patients (0.04%) required neurosurgical intervention. Patient 1 was born with transposition of the great arteries with ventricular septal defect and aortic arch narrowing. He underwent palliative Rashkind balloon septostomy on day one of life, and an arterial switch procedure on day 41, requiring extracorporeal membrane oxygenation (ECMO) after surgery. He developed an acute pan-hemispheric left subdural hematoma that required a left coronal burr hole and a subdural drain. The patient was separated from ECMO on postoperative day 4 and delayed sternal closure on postoperative day 6. The patent is now 7 years old and without significant neurological deficit. Patient 2 is a 3 year old who had undergone Norwood and Glenn procedures for hypoplastic left heart syndrome. She underwent an extracardiac Fontan operation complicated by postoperative bleeding. She developed a right holohemispheric infarct with midline shift and effacement of the ventricles. She underwent right frontotemporoparietal decompressive craniectomy. She survived with significant neurological deficits.
Conclusion: The need for neurosurgical intervention after pediatric and congenital cardiac surgery is quite rare; however, when indicated, it can offer the possibility of satisfactory outcome.

P2681 - CURRENT OUTCOME OF VENO ARTERIAL EXTRACORPOREAL MEMBRANE OXYGENTATION IN NEONATES AND INFANTS USING MAGNETIC LEVITATION CENTRIFUGAL PUMPS

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Background: Outcomes of first generation centrifugal pumps has been controversial, thus latest generation pumps using magnetic levitation technology were developed in order to overcome mechanical properties of first generation devices.
Objective: This retrospective study aimed to assess the safety and efficacy of V-A ECMO for cardiac indications in neonates and infants using standard (SP) and latest generation magnetic levitation (ML) centrifugal pumps.
Methods: Between September 2002 and January 2017, 40 consecutive patients with a median age of 23 days (4 days-12 months) and a median body weight of 3.2 kg (1.9-12; 11/40 < 2.5 kg) were supported using V-A ECMO. Indication to V-A ECMO was circulatory collapse in ICU in 18 (45%), failure to wean after cardiac repair in 12 (30%), myocarditis in 4 (10%), sepsis in 3 (7.5%) and refractory arrhythmias in 3 (7.5%). Central cannulation was used in 32 (80%) patients. Seven patients were supported with SP and 33 with ML pumps.
Results: Median duration of support was 72 hours (range 24-672), with an overall weaning rate of 82% (33/40). Overall survival to discharge was 48% (19/40). Risk factors for hospital mortality included lower (<2.5 kg) body weight ($p = 0.02$) and rescue ECMO after cardiac repair ($p = 0.03$). During a median follow-up of 52 months (range 1-97), there were 4 (10%) late deaths and 2 late survivors with neurological sequelae. Comparison of weaning rate (5/7 versus 28/33, p = NS) and prevalence of renal failure requiring dialysis (4/7 versus 16/33, p = NS) were comparable between SP and ML ECMO groups. Patients supported with ML had a trend toward higher hospital survival (1/7 versus 18/33, $p = 0.05$) and significantly higher late survival (0/7 versus 15/33, $p = 0.02$).
Conclusions: The present experience shows that V-A ECMO for cardiac indications using latest generation centrifugal pumps may further improve end-organ recovery, hospital and late survival, even in very low body weight newborn.

P2684 - TOWARDS FAST TRACKING PAEDIATRIC CARDIAC SURGERY STRATEGY AND INITIAL EXPERIENCE WITH EARLY EXTUBATION

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Objective: The aim of this study is to report our initial experience with early extubation (<6 hours)following congenital cardiac surgery, assessing its efficacy and safety and the potential for fast tracking through Paediatric Intensive Care Unit (PICU).
Methods: Early extubation was defined as intraoperative or within 6 hours from arrival to PICU. Between January 2014 till March 2016, 847 patients underwent congenital cardiac surgery at Alder hey Childrens Hospital with a 30 day mortality rate of 0.9 percent. The clinical records of 525 patients older than 90 days of age were reviewed. The mean age and weight was 1.04 years and 12.1 kg respectively. Redo-sternotomy accounted for 131 cases (25%). The management strategy involves a specific anaesthetic technique, warm cardiopulmonary bypass and intraoperative echocardiogram for evaluation of surgical repair.
Results: Out of 525 patients, early extubation was accomplished in 465 patients (88%)with an intraoperative extubation rate of (66%).

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There was no mortality or other adverse events related to early extubation. Reintubation was required in 3 patients (0.6%). Patients extubated earlier had a shorter PICU stay (3.1 + /-3.6 vs 5.1 + /- 2.7 days) and shorter hospital stays (5.9 + /- 10.7 days). It was noted that the PICU stay was artificially longer due to the bottle - neck effect along the patient flow.

P2686 - INITIAL CLINICAL MANIFESTATIONS AND RESULTS AFTER SURGICAL REPAIR OF DOUBLE CHAMBERED RIGHT VENTRICLE IN CHILDREN AND ADULTS

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Background/Hypothesis: Double-chambered right ventricle (DCRV) is a rare congenital heart disease, characterized by division of the right ventricle by hypertrophy of the septoparietal trabeculations into two parts, the proximal chamber with higher pressure, and the distal chamber with low pressure. We aim to report our experiences regarding the DCRV symptomatic presentations, long-term prognosis, including the recurrent rate of DCRV and occurrence of arrhythmias after surgery.
Materials and Methods: We retrospectively investigated 89 consecutive patients who were diagnosed with DCRV and underwent a surgical intervention from 1995 to 2016. We investigated the following clinical characteristics before and after surgery: gender; age at the time of surgery; follow-up period; pressure data obtained using transthoracic echocardiography and cardiac catheterization; and co-existing conditions, such as ventricular septal defect (VSD), atrial septal defect (ASD). The surgical approaches, postoperative mortality rate, and occurrence of cardiac events were also evaluated.
Results: Mean age at the time of diagnosis was 1.96 ± 4.15 years (1 month - 30 years) and 5.3 ± 4.89 years (5 months - 30 years) at the time of operation. Concomitant cardiac anomalies; perimembranous VSD (78 patients), musculer VSD (2 patients), ASD (9 patients), right arcus aorta (3 patients), left superior vena cava (2 patients), patent ductus arteriosus (1 patient), peripheral pulmonary stenosis (1 patient). The mean follow-up period was 4.86 ± 4.6 years. In these patients, the systolic pressure gradient in the RV by echocardiography before, immediately, and long-term after surgical intervention was 49 ± 31, 11.8 ± 7.9 and 10.4 ± 6.9 mm Hg respectively. There were no deaths during the long-term follow-up period. Surgical reinterventions were performed for residual VSD (2 patients), residual pulmonary stenosis (1 patient), severe tricuspid insufficiency (1 patient).
Conclusion: The surgical outcomes and postoperative prognosis of DCRV are favorable, and neither recurrence of DCRV nor fatal arrhythmias develop during the long-term follow-up period.

P2687 - PSYCHOSOCIAL CONDITIONS AND QUALITY OF LIFE OF PATIENTS AFTER STAGED OPERATION FOR HYPOPLASTIC LEFT HEART SYNDROME AND THEIR FAMILIES ONE CENTER EXPERIENCE

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Background: Improvement in results of staged palliation for HLHS indicates assessment of HLHS survivors quality of life (QoL).
Aim: To assess the QoL of patients with HLHS and their families.
Material and Methods: 98 mothers of patients with HLHS (73 boys – 75%) completed the questionnaire to assess their children QoL and impact of child's illness on the family. All children (age 3-17 years, mean 7 years) were operated at our institution. Prenatal diagnosis of HLHS was established in 43 (44%). Mean mother's age at the delivery was 28 years (17-41 years). In 52% child with HLHS was the first, in 28% - the only child.
Results: Development estimated as normal was reported in 89%. Limited physical activity in 80%, emotional problems in 24%, educational in 9%. Normal schools and kindergartens attendance in 78%. Learning progress estimated as good and very good in 70%. Child's illness indicated strong parental stress (93%), but 87% of responders estimated that they successfully cope with the problem. Only 11% of mothers looked for psychological care. In 94% familial atmosphere was assessed as good, in 78% child's illness strengthened parental marriage. In 18% of families mother was the only parent. Impact of child's illness on material situation was significantly negative in 79%. In 57% father was the only working parent. 97% of responders wouldn't change anything in their choice of the method and place of treatment.
Conclusions: 1. Patients with HLHS are active members of the society, they attend to normal schools and kindergartens although their physical activity is limited. 2. The family functioning is good but child's illness is a reason of strong parental stress and indicates material problems. 3. Increasing number of HLHS survivors indicates the need for continuation studies concerning neurodevelopmental outcome, quality of life, career and family planning and social functioning.

P2695 - COMPUTATIONAL FLUID DYNAMICS ANALYSIS PULMONARY BLOOD FLOW AFTER TOF REPAIR

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Objectives: We hypothesize that patient-specific computed tomography (CT)-based morphometry with TOF repair followed by reconstruction of viable surgical options with hemodynamic function assessment using computational fluid dynamics (CFD) can guide surgical decisions.
Methods: Two TOF patients (Model 1 and 2) who underwent successful correction by one pericardial patch (PP) was selected as a reference for morphological characterization by 3D model of patient-specific pulmonary artery reconstruction from CT images. The third patient (VR) with residual left pulmonary artery (LPA) stenosis after repair, was selected for virtual re-correction LPA using the same strategy (i.e. PP) by computer-aided design. CFD was employed for hemodynamic analysis of pulmonary blood flow, including of streamlines, wall shear stress (WSS), energy loss and ratio of flow distribution.
Results: Compared with patient after VR, before one showed more vortex formation in the LPA, swirling flow (1.5 m/sec vs 0.2 m/esc), higher WSS area and energy loss of LPA blood flow, with lower distribution ratio. In the VR, the streamlines from

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main PA to LPA was smoothly with more than rectangular position between main PA and LPA.
Conclusions: In patient-specific virtual re-correction LPA surgery followed by CFD evaluation has the potential in augmenting morphology-guided decisions on surgical strategy. Considering the limitations relate to the small number of patients, these data may provoke reconsidering the optimal surgical design to TOF repair.

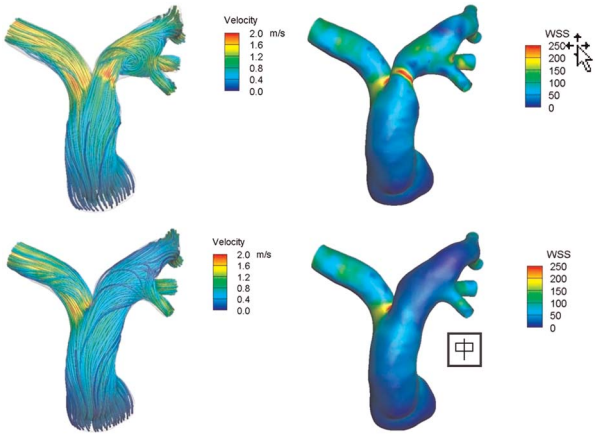

Figure.

P2702 - THE LEFT ANTERIOR MINI THORACOTOMY APPROACH FOR THE CLOSURE OF PATENT DUCTUS ARTERIOSUS IN PRE TERM NEONATES MID TERM RESULTS

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Objectives: We hereby present the mid-term results of our series, using the left anterior mini-thoracotomy technique for closure of a patent ductus arteriosus (PDA) in pre-term neonates. Left anterior mini-thoracotomy is a surgical procedure that consists of an anterior mini-thoracotomy (~1.5 cm) below the clavicle, entering the thoracic cavity from the 2nd or 3rd intercostal space, clipping the PDA, and closing the thorax without a tube thoracostomy.
Methods: Between 2009 and 2017, we performed PDA closure in 88 pre-term neonates using this method. Logistic regression analysis of potential risk factors for mortality was determined.
Results: The mean weight of the patients at the time of operation was 801.49 ± 22.03(range 450g-1500g). The mean age at the operation was 26.44 ± 3.15 days. 1 surgery-related mortality occurred and it was due to respiratory and ventilatory difficulties. Ten short-term mortalities occurred after the operation (11.3%) before the patients could be discharged from newborn intensive care unit. Five were due to sepsis, two was due to necrotizing enterocolitis and 3 were due to multi-organ failure. According to the linear regression analysis, no other variables were found to be statistically significant for predicting mortality. A negative linear correlation was found between the weight of the patients at the operation and extubation time.
Conclusion: The surgical outcome of left anterior mini-thoracotomy for PDA closure in neonates is comparable to other methods with satisfactory results. This technique may be advantageous for extremely low birth weight infants, since it causes less traction and trauma on the congested lungs.

P2706 - PREVALENCE OF ARRHYTHMIAS IN POSTOPERATIVE PERIOD OF JATENE OPERATION FOR TGA

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Background: Jatene operation is considered the treatment of choice for most TGA anatomies; improviment in mortality and less long-term complications is becoming the usual evolution in most groups. Usually arrhythmias are associated with residual lesions or coronary arteries abnormalities.
Objective: To evaluate the prevalence and caractheristics of arrhythmias in postoperative period of Jatene operation for TGA.
Material and Methods: Retrospective study included 109 patients from a single center with complete follow-up (1 to 39 years), who underwent Jatene operation (from 1976 to 2015) for TGA (65 intact septum, 29 VSD, 9 subaortic or pulmonary stenosis).
Results: 17 (15.6%) presented arrhythmia, being AV block with pacemaker implant (9), supraventricular or ventricular tachycardia (7) and atrial flutter (1). A lower incidence of arrhythmias in patients with TGA with intact septum (9.2%), compared to other complex types (26.3%) (P = 0.025). Older age at the operation was related with higher arrhythmias prevalence (p = 0.03). Cardiac failure was more prevalent in patients with arrhythmias (29.4%), compared to patients without arrhythmia (7.6%) (P = 0.03). There was no relation between the prevalence of arrhythmia and the presence of residual lesions or coronary anomaly.
Conclusion: The development of arrhythmias was associated with more complex subtypes of TGA, older age at the time of the operation and was not associated with the presence of residual lesions or coronary anomalies.

P2709 - THE EARLY OUTCOME OF DELAYED CHEST CLOSURE AFTER COMPLEX PEDIATRIC CARDIAC SURGERY – REVIEW OF SINGLE CENTRE EXPERIENCE FROM SOUTHERN INDIA

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Background: Open chest followed by delayed chest closure is a standard of care in major centers after complex cardiac surgery in the pediatric age group. The Morbidity and Mortality in our center is examined.
Methods: Between July 2014 - December 2016, 545 children with age less than 18 yrs underwent cardiac surgery. Delayed chest closure was performed in 51 children (9.3%). The mean age and weight was 231 ± 6.6 days and 4.4 ±3.17 respectively. Male predominates (72%). Cardiac surgeries included Arterial switch operation in 16 pts (31.3%), TAPVC repair in 15 (29.4%) total intracardiac repair for TOF in 6 pts (11.7%),and others in 14pts (27.4%). The Mean CPB time was 176.58 ± 78 minutes and cross

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clamp time was 102.39 ± 52.86 minutes. The chest was electively left open in 20pts (39%), for bleeding in 12(23%), long pump run in 6(11%), high PA pressure in 6 (11%),unsatisfactory haemodynamics in 4(7%)and for ECMO in 3(5%).
Results: The Mean number of days of open chest was 2.2 ±2.04. The mean number of re exploration in the ICU was 0.72 ±1.01. The mean number of Ventilation, ICU stay, antibiotic use and Hospital stay was 87.48 ±73 hours, 5.3 ± 3.8 days, 4.7 ± 3.14 days and 9.16 ±5.4 days respectively. Among these patients, 30 day mortality was observed in 22 pts (43%). Sepsis was the major cause of death (n = 8, 36.3%), followed by PAH crisis (n = 6, 27.2%) and cardiac failure (n = 5, 22.7%). The median Followup was 8 months (1 month – 30months). No late mortality.Only two patients (3.9%) developed deep mediastinal wound infection on followup and needed surgical intervention.
Conclusion: Delayed chest closure effectively optimizes the cardiac status after complex cardiac surgery in children.However delayed chest closure is associated with high postoperative sepsis and there by increases the 30 days mortality.Therefore judicious use of delayed chest closure is recommended.

P2714 - POVERTY & PERCEPTION OF HEALTH IN INTERNATIONAL PEDIATRIC CARDIAC SURGERY PATIENTS

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Introduction: CardioStart's international compassionate missions are structured to provide intensive teaching and equipment to support local congenital and acquired heart surgery program development. Determining how pediatric patients are selected and whether or not preventative care issues can be effectively addressed, is often uncertain.
Methods: Data from patients awaiting cardiac surgery by our visiting teams was examined in 13 missions to evolving tertiary care hospital centers in 8 countries, and in-depth direct interviews of 78 pediatric patients or their immediate accompanying relative carried out. Documentation included full cardiological evaluation and questions regarding patient's living conditions, time from first cardiac diagnosis until surgery, and their perception of current state of their health. Diagnoses included Atrial Septal Defect (18), Mitral Valve disease (4), Fallot's Tetralogy (8), Ventricular Septal Defect (30), Patent Ductus Arteriosus (10), and others (8). Patients' socio-economic and poverty status were identified by using typically used poverty indicators (including water obtained by well only, dirt floor, living in temporary shelters, electricity supply and food scarcity measures).
Results: Those identified as [1] 'extremely poor' reported lower perceptions of health when compared to other pediatric patients in the same geographic area [2], with similar diagnoses (2.14 vs 3.18 [3], p = .005). Gender and time from diagnosis until surgery were not correlated with differences in perceptions of health. Patient's poverty level did not correlate with longer waiting time between first cardiac diagnosis and surgery.
Conclusions: Mission locations have diverse cultural and medical specialist access issues: the lack of gender differences, and the comparable evaluation and referral times are encouraging. Our international missions already include structured Outreach rural clinic assistance. These Poverty and Health Perception results, however, underscore the need for emphasizing primary, preventative cardiac disease related education. This may help strengthen primary and tertiary cardiac care interaction [4].

P2731 - SURGERY FOR LEFT VENTRICULAR OUTFLOW TRACT OBSTRUCTION IN NEONATES AND INFANTS

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Background: Infants and neonates with severe left ventricular outflow tract obstruction can be treated with pulmonary autograft replacement of the aortic root. In this retrospective single center cohort study, we present our experience with the Ross(-Konno) procedure in neonates and infants with a focus on survival, durability of the autograft and homograft, and left ventricular function.
Materials and Methods: Retrospective observational study of 23 infants (age <1 month (n = 8), <1 year (n = 15)) operated on in the Wilhelmina Children's Hospital, Utrecht, the Netherlands between 2004 and 2016. Echocardiography was used to determine left ventricular function in the post-operative follow-up.
Results: Patients had a pulmonary autograft replacement of the aortic valve with (17) or without (6) septal myectomy (Konno-procedure. Concomitant coarctectomy (5), correction of an interrupted aortic arch (6) and resection of endocardial fibroelastosis (6) was additionally performed. Median patient age at surgery was 72 days (5–342 days). All patient were available for follow-up. Median follow-up was 4 years (0,5-12 years). Overall mortality was (1) 5,9%. Freedom from autograft reintervention was 100%. Echocardiography demonstrated good LV function, even in poor contracting chambers with endocardial fibroelastosis at the time of initial surgery.
Conclusion: A Ross(-Konno) procedure can be performed with good results in neonates and infants for treatment of severe LVOT obstruction. The pulmonary autograft is a durable substitute. This procedure offers an effective desobstruction of the LV and may allow LV function to normalize.

P2735 - AN ALGORITHM FOR SURVIVAL OF THE CHALLENGING POSTOPERATIVE LATE PRESENTING FONTAN PATIENTS

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Objective: We hereby present our case series for a group of late presenting fontan patients, which had a challenging early postoperative course.
Methods: In the last 2 year period, we dealt with 6 late presenting fontan patients with a misleading postoperative course, age ranging between 6 to 18 years. Uneventful classic extracardiac fontan

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procedures end-up with signs of a downhill multiorgan failure phase in the intensive care unit(ICU). We immediately applied an A-V extracorporeal membrane oxygenation system (ECMO) to stay one step forward, before evaluating the underlying cause. After the patients were stabilized hemodinamically, they all underwent cardiac catheterization under ECMO to evaluate the reason for the instability.
Results: All of the patients underwent ECMO for initial stabilization and in 1-2 days to cardiac catheterization in the cathlab for evaluation. According to the findings, the patients have either a surgical reconstruction or an interventional procedure like balloon dilatation of the fenestration and/or a dilatation or stenting of the pulmonary arteries. 2 of the patients were discharged uneventfully after almost 3 months in the ICU, 2 of them are still in the ICU (45 days and 3 months), and 2 of the patients did not survive because of multiorgan failure.
Conclusion: The late presenting Fontan patients are difficult to manage and needs the medical staff to think one step ahead for satisfactory results. We believe that when the patient is showing signs of multiorgan failure, acting immediately is mandatory, ECMO is essential and protecting the circulation of the endorgans with ECMO from the complications of cardiac catheterization and contrast dye is also important. We also prefer to use a side graft as chimney to the extracariac fontan conduit for venous cannulation of the ECMO, which not only helps with the uneventful drainage to the circuit, but also facilitates the interventions in the cathlab.

P2737 - OPTICAL TOOL TRACKING FOR SKILL ASSESSMENT IN OPEN SURGERY

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Background/Hypothesis: Optical tool tip tracking for surgical skill assessment can be effectively performed on videos captured via mobile phones and processed using computer vision methods. By utilising an affordable and ubiquitous device for filming, the technology can be used for skill assessment without requiring an external observer.
Materials and Methods: An operator was set up to perform suturing on a synthetic suturing pad while wearing surgical gloves with specially designed chequerboard patterns and using tools (needle holder and forceps) modified with colour markers. The camera was first calibrated and then used to film the operator as they sutured. The videos were processed by detecting the chequerboards and using the Perspective-n-Point to recover their 3D orientation. From that, a kinematic chain was established from the chequerboards to the coloured markers on the tips of the tools, resulting in 3D positions and orientations for the tool tips.
Results: Validation results using a commercial electromagnetic tracking system showed that the hands could be tracked effectively using computer vision methods on mobile phone video. Further improvement is required on the kinematic chain due to variability in holding tools and movement during suturing.
Conclusions: A mobile phone can be used to effectively track the 3D positions and orientations of the hands during suturing. Our next steps are to improve the method to work in any lighting condition and to incorporate the developed framework into a mobile application for medical students to assess their suturing and knot tying skills.

P2739 - PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM EARLY MANAGEMENT AND MID TERM OUTCOMES

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Background: Pulmonary atresia with intact ventricular septum (PAIVS) is a complex and uncommon congenital heart disease, with a wide morphologic spectrum. The aim of this study was to describe the surgical management and results according to the different anatomical variants.
Methods: Between 2000-2015, 67 patients (p) with PAIVS were operated on, with a median follow-up of 7,92 (IQRR 5-11-5 years) and 10p were lost in follow-up. Membranous atresia occurred in 81% of the patients. Right ventricular dependent coronary circulation was observed in 13% and tricuspid valve dysplasia in 14%.
Results: A shunt was performed in 54p (80%) at a median age of 17 days with an early mortality rate of 9.2% (5p, three of them with concomitant right ventricular outflow tract obstruction relief). It was indicated a biventricular approach as initial strategy in 30p: 16p (53%) reached a total biventricular stage and 14p (47%) partial repair. In 27p the final stage was univentricular pathway, at a median age of 5 years, 3p are awaiting for complete repair. The table 1 summarizes the morphologycal characteristics and different procedures.
Conclusions: Thorough evaluation of the morphology is essential to define the strategy of management. A favourable initial tricuspid annulus size (Z score > -3) was associated with a final biventricular repair, either total or partial. In contrast, bipartite right ventricle morphology was not related to final stage. Early right ventricular outflow tract obstruction relief allowed the possibility of a biventricular pathway. The choice of initial management and better selection of patients allows planning biventricular repair with good outcomes, despite the fact that it requires multiple reinterventions. Univentricular repair had larger mortality rate. Nevertheless, the three groups showed similar functional capacity in the mid-term follow up..

Table.

	Biventricular (16p) 28%	**One and a half (14p) 25%**	**Univentricular (27p) 47%**
Tricuspide Valve Z score			
>-3	85%	70%	
<-3			90%
RV: tripartite	56%	50%	
RV: bipartite	44%	50%	68%
RV: unipartite			32%
RV decompression:			
Transcatheter	6p (37.5%)	6p (43%)	
radiofrequency (RF)	Median age: 30 days	Median age: 142 days	
Surgery	10p (62.5%)	8p (57%)	
	4p valvotomy	2p transjunctional patch	
	5p Homograft	6p valvotomy	
	1p transjunctional patch	Median age 13 days	
	Median age 37.5 days		
RV overhaul	5	6	
Tricuspid valve repair	7	3	
Mortality	0	1p (7%)	4p (15%)
NYHA grading			
Class I	85%	40%	62%
Class II	15%	60%	38%
Reinterventions	10p (62,5%)	8p (57%)	7p (26%)

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P2741 - AUTOLOGOUS PULOMONARY ARTERY TUBE RECONSTRUCTION TO ESTABLISH DUAL CORONARY SYSTEM IN ALCAPA FROM NON FACING SINUS

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Background: Various techniques have been described for establishing dual coronary system in ALCAPA repair. But management of ALCAPA from posterior non facing sinus is more challenging. We present our modest experience with pulmonary artery tube reconstruction in this condition.
Materials & Methods: From Jan 2013 to Sep 2016, eleven babies underwent ALCAPA in our institute. Out of 11, five babies had LMCA arising from posterior non facing sinus.Our study included only these five patients. Male : female ratio was 1:4. Age at presentation varied from 23 days to 7 months. All babies underwent ALCAPA repair under cardiopulmonary bypass using aortic and bicaval cannulation.
Results: Dual coronary system was created using autologous pulmonary artery tube in all the five babies. After harvesting LMCA from the pulmonary artery ,a pulmonary artery tube was created by suturing the pulmonary artery wall of the sinus portion. The mean CPB time and aortic cross clamp time were 179 minutes and 77 minutes respectively.The chest was left open in two babies.We had one postoperative death due to persistent low cardiac output. ECMO support was used in one patient.Left phrenic nerve palsy was noted in one bay who underwent diaphragmatic plication.
Conclusion: From our experience, we conclude that pulmonary artery flap tube repair of ALCAPA helps in creation of a long, tension free neo LMCA and perfect aortic reimplantation to establish the dual coronary system in babies with ALCAPA from non facing sinus.

P2751 - DUCTAL ASSOCIATED PULMONARY ARTERY STENOSIS (PULMONARY COARCTATION) WITH NEONATAL BLALOCK TAUSSIG SHUNT INCIDENCE AND MANAGEMENT

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Background: Infants undergoing a modified Blalock-Taussig shunt (MBTS) can have stenosis at the ductal insertion site on the pulmonary artery (PA) termed pulmonary coarctation. We examined our experience with pulmonary coarctation at our institution.
Patients and Methods: From 2002, 311 infants aged <90d (0-87 days, mean 17.4 ± 21.2; weight 1.4-6.2 kg, mean 3.1 ± 0.7) had a primary MBTS; 161 (52%) had univentricular anatomy. Four diagnoses each with >10% of the total number of shunts accounted for 59% of the shunts: pulmonary atresia with intact ventricular septum (n = 65, 21%), pulmonary atresia with ventricular septal defect (PA/VSD; n = 47, 15%), tricuspid atresia (n = 38, 12%) and tetralogy of Fallot (n = 33, 11%). Pulmonary coarctation was identified by preoperative echocardiogram or catheterization or at direct intraoperative inspection and repaired at the time of the MBTS.
Results: 73 infants (23%) had pulmonary coarctation repair with the MBTS, 61 (86%) without cardiopulmonary bypass (Figure). Patients with PA/VSD or double-inlet left ventricle (DILV) had an increased incidence of pulmonary coarctation. Pulmonary coarctation patients were younger (11.2 ± 15.1d vs. 19.8 ± 22.7d); hospital mortality was not different. Sixteen (5.1%) had late recognition of pulmonary coarctation. 26 of 89 pulmonary coarctation patients (29%) had recurrent stenosis including 5 of 6 with situs inversus and right pulmonary arterioplasty.
Conclusions: Pulmonary coarctation is common in infants undergoing MBTS, especially with PA/VSD or DILV. It usually can be managed without CPB during the initial MBTS. Operative mortality is not different from patients with MBTS alone. Delayed recognition and late recurrence after pulmonary arterioplasty are not infrequent; ongoing vigilance is necessary.

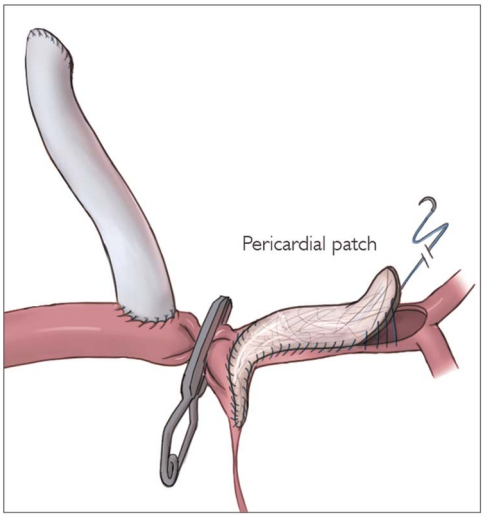

Figure.

P2758 - LEFT VENTRICULAR REMODELING AT ONE YEAR AFTER AORTIC VALVE REPAIR IN PATIENTS WITH CONGENITAL ISOLATED AORTIC REGURGITATION

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Background: Congenital isolated aortic regurgitation (AR) is a rare congenital defect in the young. Chronic left ventricular (LV) volume overload results in irreversible myocardial dysfunction. We aimed to examine changes in aortic valve and LV size and function from baseline prior to aortic valve repair (AVre) to discharge and 1 year follow-up after.
Methods: Single institution, retrospective review of subjects <25 years who underwent AVre initially for isolated AR between 1989 and 2014. Inclusion criteria were $\geq$ moderate AR and mean Doppler gradient ≤ 30 mmHg at baseline (BL). Aortic valve annular, aorta ascendance and LV size and LV functional echo variable Z-scores at BL, discharge (DC) and 1 year after AVre were evaluated. The AR was assessed by vena contracta diameter adjusted to body surface area.
Results: Of 47 subjects, 39 (79.5%) underwent repair. Median age was 14 years (range 2, 24). Patients with bi- (40%) and tri- (38%) commissural AV have a greater proportion of severe AR than those with unicommissural (21%, P = 0.08). At DC, AR decreased

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to mild or less in the majority of patients (87%), resulting in significant acute decline in LV end-diastolic size and volumes, while LV mass didn't change (Table 1). The acute fall in preload resulted in increased LV mass/volume ratio and decreased ejection fraction (EF) at DC (both $P < 0.001$). At 1 year follow-up, LV end-diastolic and stroke volumes significantly increased due to AR recurrence, at least moderate grade in 62% of patients. Regardless, the LV mass significantly decreased, resulting in mass/volume ratio and EF normalization.

Conclusion: Moderate and severe AR declined to mild after aortic valve surgery, accompanied by acute LV preload reduction in the majority of patients at DC. Acute decline in LV pre-load led to ejection fraction deterioration early after aortic valve surgery, with full recovery after 1 year follow-up.

Table 1. Aortic Valve and LV Size and Function at Baseline, Discharge, and 1 Year Follow-up after Aortic Valve Surgery

Aortic Valve Morphology	Baseline (median 34 days)	Discharge (median 3 days)	1 Year Follow-up (median 365 days)	P§ Value
AR Grade (n = 45,45,26) None to mild Moderate to severe	0 (0%) 45 (100%)	39 (87%)★ 6 (13%)	10 (38%)★★ 16 (62%)	<0.001
LV EDV z-score (n = 39,29,20)	5.24 ± 1.77	2.27 ± 2.11★	2.95 ± 3.74★	<0.001
LV SV z-score (n = 39,28,19)	5.36 ±2.38	0.78 ± 2.23★	2.67 ± 3.14★#	<0.001
LV EF z-score (n = 39,38,22)	-1.03 ± 1.79	-2.91 ± 2.21★	-1.59 ± 2.10	<0.001
LV mass z-score (n = 35,26,18)	4.56 ± 2.24	4.36 ± 2.19	2.60 ± 2.57★	0.02
LV mass/ volume z-score (n = 35,34,21)	-0.81 ± 1.67	1.96 ± 2.65★	-0.11 ± 1.65★	<0.001

★- *Significantly different from BL vs. DC;#-Significantly different from DC to 1 year after surgery,§ - ANOVA comparison*

P2759 - TRACKING MORTALITY IN PEDIATRIC PATIENTS WHO RECEIVE OPERATIONS BY VISITING CARDIAC SURGICAL TEAMS

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Introduction: Between 2011 and 2016, CardioStart International teams taught and performed pediatric and adult cardiac surgeries for patients in 8 severely healthcare restricted locations, addressing the poorest within the local communities. Local teams often took the primary surgical role to provide surgeries to those who would otherwise go without. Difficulties in obtaining post-operative tracking information of 30-day, and later morbidity/mortalitY estimations are frustrated by: • Lack of electronic medical record capability; • Similarity in family names;.

• Incomplete demographic information at admission; • Inadequate and varied access to patients following discharge; • Very long traveling distances between referral center and patient's home; • Lack of primary care support; • Non-attendance to specialist follow-up clinics; • A substantial fear that follow-up attendance will incur additional costs to the patient/family, • Non-permanent addresses, shared housing and/or internal migration by families for work. • Surgeons/cardiologists leaving one tertiary care center for better opportunities elsewhere;.

Methods: We successfully tracked 78 pediatric patients who underwent cardiac surgery between 2011 and 2016, in 13 missions to Brazil, Myanmar, Vietnam, Dominican Republic, Peru, India, Nigeria, and Uganda. Information regarding live status and well-being were obtained by contact with specialist doctors and patients through e mail and cellphone using a RedCap-based questionnaire.★.

Results: At 30 days, 70 patients were confirmed as alive, 5 were confirmed as mortalities, and only 3 remained unknown, confirming a successful tracking rate of 96%.

Conclusions: Tracking of patients in start-up programs or those centers seeking skill advancement is difficult in limited resource environments, but does remain possible. A significant improvement in tracking success was obtained by clearly stating to patients on discharge that no financial charges for follow-up information received would be made to them on any repeat evaluations. Connectivity through E mail and cellphone (direct/indirect through relatives) was surprisingly high. ★(Bio-Statistic Function of the Clinical and Translational Science Institute, University of Minnesota).

P2770 - NEW BIOSCAFFOLD USED FOR SURGICAL REPAIR OF COARCTATION OF THE AORTA IN NEONATES

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Background: In neonates with a low body weight with long segmental coarctation of the aorta (CoA), plastic repair technique might be useful. CoA commonly coexists which other heart defects, e.g. ventricular septal defect (VSD) or complex heart disease. To surgically correct CoA, surgeons can use several biomaterials. The newest is a decellularized bovine pericardial patch. The aim of the study is to present the results of CoA repair with the decellularized bovine pericardial patch.

Material and Methods: We retrospectively analyzed 11 neonates with coarctation of the aorta, in which the decellularized bovine pericardial patch was used to repair the defect. All the procedures were performed through the postero-lateral thoracotomy. The follow-up examination included echocardiography and clinical examination.

Results: The mean patient age was 9 days (range 4–116); mean body weight 3.3 kg (range 1.4–10.8). Nine patients (81%) had other heart defects, mainly presenting with VSD (nine patients), followed by transposition of the great arteries (2), aortic arch hypoplasia (2) and BAV (2). The mean ICU stay was 7 days (range 4–13). The mean time from surgery to follow-up was 7 months (range 4–13). Recoarctation of the aorta was found in three cases

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(27%), 4, 6 and 8 months after surgery. In echocardiography, the maximum gradient through CoA was in the range of 33-80 mmHg (mean - about 50 mmHg). These patients had balloon plasty and one of them had hypoplastic aortic arch. Balloon plasty was effective in all the patients. Only two of our patients were treated with aspirin. None of them had any problems with recoarctation.

Conclusion: A longer follow-up is needed to assess the usefulness of the decellularized bovine pericardial patch for aortic surgical repair in neonates.

P2774 - PROSPECTIVE ASSESSMENT OF STERNOTOMY WOUND RELATED COMPLICATIONS AFTER PAEDIATRIC CARDIAC SURGERY

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Background: Many previous studies of wound-related complications post-paediatric cardiac surgery have focused primarily on unmodifiable risk factors or have used definitions non-specific to the sternal wound. This study aimed to prospectively identify the true incidence of infective and non-infective wound complications, providing a tool for comparison and monitoring of such complications.

Methods: A prospective 90-day surveillance programme was developed to identify all sternotomy wounds that failed to heal by primary intention in which at least one of the following were present: wound collections /dehiscence, abnormal or persistent inflammatory signs/symptoms with or without positive inflammatory markers or microbiology cultures. Sternum and/or mediastinum involvement placed the complication in the deep group. Frequently-cited associated variables were analysed and cumulative sum control charts (CUSUM) were derived for monitoring deviations from target values in real time. The impact of a wound care bundle on outcomes was assessed over a 2-year period.

Results: There were 105 wound-related complications in 1020 sternotomies with 88(8.63%) categorised as superficial and 17 (1.66%) as deep. These required 362 surgical wound reviews, and 58 surgical interventions (Table1). Only 13(14.4%) of superficial wounds elicited positive cultures whereas 14(82.35%) did so in the deep group, mostly Staphylococcus species (Table1). More than half of the complications in the deep group initially manifested superficially. Demographic variables associated with wound infections are displayed in table. The implementation of a new wound care bundle demonstrated a significant impact on the incidence of superficial wound complications, but not on those classified as deep.

Conclusions: A real-time surveillance programme of sternotomy wound complications provides a more comprehensive assessment of the true incidence of infective and non-infective delays in healing. This represents an important quality metric that informs resource allocation and allows long-term performance monitoring. The implementation of a wound care bundle reduced superficial wound complications but not those involving the sternum and/or mediastinum..

Table 1. All values are presented as median and IQR except for RACHS cotegaries (%) and Management, Neonotes, First sternotomy, and Positive culture (absolute values). Of note, 25% of the wound complications were diagnosed beyond the 3rd quortile

Variable	All sternotomies (n = 1020)	Superficial wound complications (n = 88)	Deep wound complications (n = 17)
RACHS category			
1	5.7	3.5	5.9
1	33.6	15.9	29.4
3	41.9	38.7	23.5
4	5.5	12.5	0.0
5	3.6	4.5	5.9
6	7.8	23.9	35.3
Unclassified	2.0	1.1	0.0
Age (days)	300 (70-1,686)	19 (4-212)	325 (6-601)
Neonates	200 (19.6%)	46 (52.2%)	6 (35.3%)
Weight (Kg)	7.6 (4.1-16.1)	3.5 (3.0-6.2)	8.2 (3.6-10.4)
CBP time (min)	77 (54-117)	106 (67-130)	118 (89-140)
PICU LOS (h)	32 (22-119)	142 (71-254)	72 (24-219)
First sternotomy	623 (61.0%)	69 (78.4%)	10 (58.8%)
Days to first review	NA	8.5 (5.8-12.3)	12.0 (10.0-17.0)
Management			
Conservative	NA	240	64
Minor surgical procedure	NA	24	10
Wound debridement + /- Vaccum therapy	NA	0	5
Wound debridement and sternal re-wiring	NA	0	19
Positive wound culture	NA	13/90 (14.4%)	14/17 (82.3%)
		-S. aureus 5 -S. coagulase-negative4 -Klebsiella pneumoniae 1 -Haemophilus influenza 1 –E. coli 1 -Candida albicans 1	S. aureus 11 S. coagulase-negative 1 S. haemolyticus 1 E. coli 1

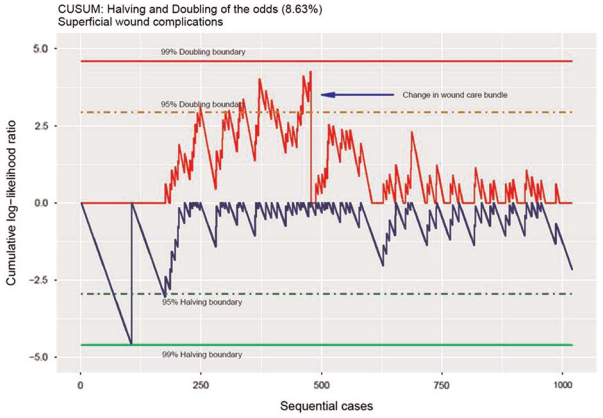

Figure 1.

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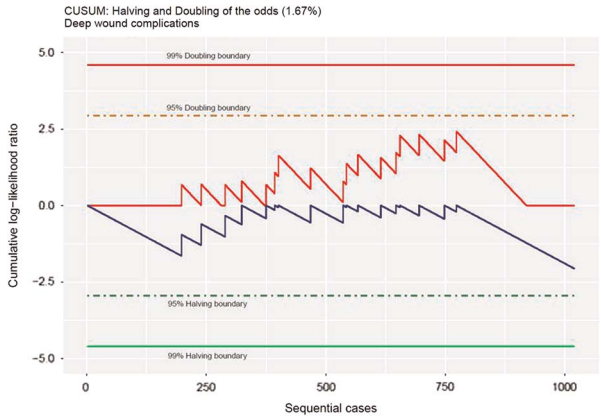

Figure 2.

P2788 - CASE REPORT TRICUSPID ATRESIA WITH COMMON ARTERIAL TRUNK FIRST STAGE SURGICAL PALLIATION

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Tricuspid atresia (TA) and common arterial trunk (CTA) are rare anomalies accounting for 2.5% and 0.7% of all congenital heart diseases, respectively. The coexistence of both anomalies is rare. This association constitutes only 0.01% to 0.02% of congenital heart disease.
Case Report: A Three-months-old female infant weighing 3.4 kg. A transthoracic echocardiogram performed at the time showed tricuspid atresia with a hypoplastic right ventricle, with truncus arteriosus type one, mild truncal valve regurgitation, an unrestrictive interatrial communication, and good-sized branch pulmonary arteries. Cardiac computerized topography (CT) angiography confirmed the diagnosis. The case was approached via median sternotomy, both pulmonary artery branches were snared, the VSD was closed using gortex patch, the right ventricular outflow was reconstructed with direct RV-PA conduit using aortic homograft size 11 which was the smallest size available, then banding of the homograft was done to get a mean pulmonary artery pressure of 25 mmHg. Pre discharge echocardiography revealed good ventricular function, moderate aortic regurgitation and mild regurgitation across the right ventricle to pulmonary artery homograft with a pressure gradient across the band 50 mmHg. The patient was discharged from the hospital in a stable condition.
Conclusion: The association of CTA and TA is an extremely rare finding and represents a particular challenge for the medical and surgical team. Without surgical palliation, the definite outcome is a neonatal death. Various approaches to the management of this morphology have been described with varying success, and consensus remains to be established. We demonstrate that successful first stage palliation of this condition can be undertaken with satisfactory results using a Sano shunt as primary palliation.

P2791 - AORTIC ARCH RECONSTRUCTION AND BILATERAL PULMONARY ARTERIES BANDING – NEW MODIFICATION OF THE NORWOOD OPERATION FOR TREATMENT OF HYPOPLASTIC LEFT HEART SYNDROME – EXPERIENCE OF OUR CENTER

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Background: In the period from 2013 to 2016, 46 Stage I procedures were performed in our center on children with hypoplastic left heart syndrome (HLHS). The study sought to retrospectively evaluate early results of a new modification of the Norwood operation for treatment of hypoplastic left heart syndrome.
Material and Methods: The group analyzed included 46 children with HLHS after modification of the Norwood procedures, in whom the aortic arch was reconstructed with an extracellular matrix patch and bilateral pulmonary artery banding.
Results: The mean patient age was 16 days (1-47 days) and the mean body mass was 3025 g (1500 to 4200 g) on the day of operation. The overall hospital mortality rate was 15.2%. Twenty-one patients were discharged directly from our department to outpatient care after an average of 18 days. The remaining 18 patients were in stable condition and transferred to a pediatric clinic for follow-up treatment after an average of 25 days of postoperative care. The rates of death were: 1 patient (14,3%) due to bleeding, 2 patients (28,6%) due to sepsis, and 4 patients (57,1%) due to low cardiac output. Eight children (39,1%) underwent open-chest management postoperatively. Postoperative bleeding requiring chest revision occurred in 8 children (17,4%). Postoperative intubation in livingpatients lasted for an average of 8 days (from 2 to 28). Assisted circulation with ECMO was employed on 5 children (10.8%). In two of these cases, assisted circulation was discontinued with a positive effect.
Conclusions: According to the assessment of results in our center the initiation of this new surgical technique enabled reduction of postoperative bleeding rates as well as shortened operation time. This method also provides better outcome in children with low birth body weight, preterm neonates and children with complicated heart defects eg. HLHS with TAPVC. There is also a need of long-term assessment of this procedure.

P2795 - SURGICAL MANAGEMENT OF ADDITIONAL MUSCULAR DEFECTS BY 'TWO PATCH TECHNIQUE' FOLLOWING EN FACE RECONSTRUCTION OF VENTRICULAR SEPTUM

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Background: Closure of additional muscular ventricular septal defects (VSD) remains a challenge. The complex anatomy of muscular septum makes it difficult to identify and address

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additional muscular VSDs. We addressed this issue by En Face reconstruction (EFR) of RV septum during pre-operative echocardiography and then a 'Two-Patch Technique' (TPT) to surgically close all additional muscular VSDs.
Materials and Methods: 39 patients (M:F = 25:14) underwent surgical repair by TPT for additional defects. All were located and mapped by pre-operative echocardiography and EFR. Mean age was 1year 9months (2 months- 10years), and mean weight 6.02 kg (2.3- 19 kg). Right-angled tip forceps was introduced through additional VSD from RV side of septum. Another forceps was introduced into LV through larger defect and a vessel loop was threaded across the defect. PTFE patch anchored to LV limb of vessel loop using 2 fine polypropylene sutures was placed on LV septum by pulling the vessel loop out through RV. Another PTFE patch was placed on RV side of defect using same polypropylene sutures- like a Hammock. Alternatively, we approached LV through inter-atrial septum across the mitral valve. Intra-operative echocardiography was done and RA-PA saturations checked. Morphological spectrum included multiple VSDs (perimembranous 36, anterior muscular 13, posterior muscular 16, apical 9, mid-muscular 9), TOF (n = 6), DORV (n = 2).
Results: Mean clamp time and bypass time was 93 mins and 14 mins respectively. Significant RA-PA step-up (n = 5) was confirmed by intraoperative echocardiography and all were banded in the same sitting. Mean hospital stay was 11.4 days with zero hospital mortality. On follow up, all patients remain in NYHA class I. All banded patients either have no residual shunt (n = 3) or tiny shunt (n = 2) awaiting debanding.
Conclusion: It is an easily reproducible technique (EFR with TPT) and can be employed for all types of additional muscular defects. Short-term results are satisfactory.

P2798 - TAKEUCHI TECHNIQUE IN FOUR CASES OF ANOMALOUS LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY

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Anomalous left coronary artery from the pulmonary artery (ALCAPA), also known as Bland-White-Garland syndrome(1) is a rare congenital cardiac disease that can cause myocardial infarction, heart failure and even death in paediatric patients(2), is present in 1 of 300 000 live births and accounts for 0.05% of congenital cardiac malformations (3). Surgical treatment is ever the treatment election and success overlive is most than 90%; coronary reimplatation is now the elective treatment when it is not possible, takeuchi technique is the elective treatment; this technique repairs intrapulmonary tunnel and is a unique procedure for repairing anomalous origin of the left coronary artery with left and posterior possition from the pulmonary artery (3)Four female clinical cases are presented: a 4 years old, 5,8 y10-months all patients with dilated cardiomyopathy with severe ventricular dysfunction, LVEF 15%, 20% 20% and and 40%, respectively, with moderate mitral insufficiency secondary to anomalous origin of the left coronary artery Of the Pulmonary Artery Trunk (ALCAPA), Takeuchi surgery was performed in all cases, with excellent results given by recovery of ventricular function as a whole. Follow-up time of 6 to 3 years..

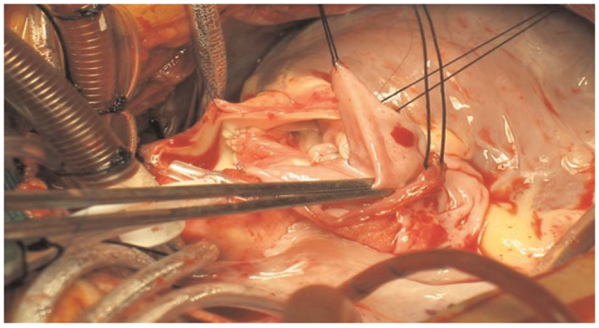

Figure 1.

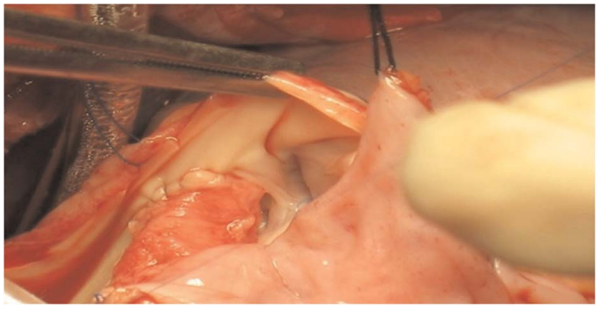

Figure 2.

Table 1. Characteristics of patients who underwent surgical repair with takeuchi technique.

Year	Age	Pre surgical LVEF	Post-surgical LVEF	Follow up LVEF (time)	Followup MI
2008	4 years old	15%	38%	65% (3 years)	Moderate
2013	8 months	20%	45%	60-70% (3 years)	Moderate
2015	5 months	20%	45%	65% (1 year)	Moderate
2016	9 months	40%	40%	65% (6 months)	Low

P2812 - CASE REPORT ORIGIN OF THE RIGHT PULMONARY ARTERY FROM THE ASCENDING AORTA

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Background: Origin of the right pulmonary artery (RPA) from the ascending aorta is an extremely rare congenital anomaly. It is anatomically different from discontinuos pulmonary arteries where one branch of the pulmonary arteries is supplied either by an arterial duct (PDA) or by major aortopulmonary collaterals. Furthermore, it is not unusual that while RPA pressure is evidently systemic, right ventricular (RV) and left pulmonary artery (LPA) pressure may be suprasystemic. The latter may reflect a discrepant degree of pulmonary vascular disease between the right and left lung increasing surgical risks along with a somewhat unpredictable outcome.
Case Presentation: A 3-year-old boy was admitted with symptoms of severe congestive heart failure. Diagnosis was non-conclusive

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on echocardiogram. A diagnostic cardiac catheterization was performed finding an anomalous RPA arising from the aorta. Concomitant defects included a small PDA and atrial septal defect. RV pressure was suprasystemic (80 vs.70 mmHg). LPA pressure was 80/35-55 mmHg compared to 70/35-47 mmHg in the RPA. Left lung wedge angiography showed more advanced stages of pulmonary vascular disease when compared to the right lung. Provided signs of congestive heart failure were still present, surgical repair was advocated. Reimplantation via an end-to-side anastomosis of the anomalous RPA into the MPA was performed using a 16mm PTFE graft. In addition, PDA closure was performed along with a lung biopsy. Systemic and main pulmonary artery pressures in the immediate postoperative period were 100/55-70 mmHg and 43/18-26 mmHg respectively. The patient had a remarkable uneventful recovery and remains under surveillance as an outpatient.
Conclusion: Anomalous origin of the right branch pulmonary artery from the ascending aorta is a rare congenital anomaly. Significant pulmonary arterial hypertension is usually present with different degree of vascular involvement between the right and left. Surgical risks are considerably higher in this population but surgical repair remains an option in selected cases.

P2817 - EARLY PRIMARY REPAIR OF TETRALOGY OF FALLOT IN EARLY INFANCY – CHALLENGES & OUTCOMES !

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Background: The timing of intra cardiac repair for Tetralogy of Fallot keeps evolving. The practice of early primary repair of TOF is recommended in babies with adequate pulmonary anatomy. We present our experience in early primary repair of TOF in infants who are less than 3 months of age.
Materials & Methods: We performed an early primary TOF correction in 41 infants who were less than 12 weeks during a period of 5 years from Sep 2011 to Aug 2016. The age ranged from 23 days to 12 weeks. Based on the preoperative ECHO, 53 patients were selected for an early repair.But, we found near pulomanry atresia and hypoplastic branch pulmonary arteries in 12 babies. Hence they underwent BT shunt and were excluded from the study group. All babies underwent intra cardiac repair with conventional cardiopulmonary bypass using an aortic and bicaval cannulation at core temperature of 28 °C.
Results: We performed transatrial repair in 29 patients and 12 had transannular patch. Seven patients needed prolonged ventilation and the mean duration of icu stay was 127 hours. There were 4 hospital deaths and one late death. Left phrenic nerve palsy was noted in 3 patients and they underwent diaphragmatic plication. The mean hospital stay was 14 days.
Conclusion: We conclude that early primary repair of TOF can be safely performed in infants less than 3 months of age with recurrent hyper cyanotic spells. Babies with good pulmonary anatomy had better clinical outcomes.Precise surgical techniques and attentive post operative care play a vital role in these babies.

P2819 - THE ROSS AND ROSS KONNO PROCEDURE IN NEONATES AND INFANTS INTERMEDIATE TERM RESULTS FUNCTION AND GROWTH OF AUTOGRAFT

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Background: The Ross procedure is commonly used in children requiring aortic valve replacement with the possibility to enlarge small aortic root (Ross-Konno). However, is not clear the potential for growth and the durability of the autograft in neonates and infants. We assess a review our intermediate-term results of the Ross and Ross-Konno procedure.
Material and Methods: A retrospective analysis of all consecutive patients aged younger than 1 year who underwent the Ross or Ross-Konno procedure was recorded. We included 13 patients (8 Ross and 5 Ross-Konno; 4 neonates, 9 infants) The median aged was 150 days and median weight was 6 kg. 11 patients had aortic stenosis, 2 patients had isolated severe aortic insufficiency and 4 patients had other left-side heart lesions, such as arch obstruction (2 patients) or mitral valve disease (2 patients). 2 patients had left ventricle endocardial fibro-elastosis. The median aortic valve diameter z-score was -1,5 (1,5 to -3,5). Major study outcomes included patient survival, postoperative complications, autograft function and need for reoperation.
Results: The median follow-up was 60 months. No patients had 2 + or more aortic insufficiency and/or moderate aortic stenosis. 4 patients required concomitant aortic arch or mitral repair. 2 patients needed ECMO after surgery. 2 patients presented renal insufficiency with depuration therapy and 2 pacemakers were implanted due to postoperative av block. No patients suffered acute myocardial infarction. Actuarial survival rate was at 100% at 5 years. We assessed an increased of autograft diameter (-1,5 to 1,7 z-score, $p < 0,003$). There were 2 reinterventions, including 2 procedures on the right-side conduit. No reinterventions was in autograft.
Conclusions: The Ross and Ross-Konno procedure for neonates and infants has good results with low mortality and acceptable rates of reintervention with potential growth.

Table 1.

Number	Age	Weight	Diagnose	Previous procedure	Preoperative z-score	LHlesions
1	116	4,1	Shone syndrome	Aortic valvuloplasty	-2,6	Mitral stenosis, fibro-elastosis LV
2	152	6	Shone syndrome	NO	-2,7	Coarctation
3	181	6,5	Aortic stenosis	Commissurotomy	1,5	NO
4	149	5,8	Aortic stenosis	Aortic valvuloplasty	-1,5	NO
5	339	12	Aortic insufficiency (endocarditis)	NO	1,57	NO
6	27	3,5	Aortic stenosis	NO	-1,2	NO
7	296	10	Aortic stenosis	Aortic valvuloplasty	-3	NO
8	237	9,5	Aortic insufficiency (endocarditis)	NO	0,8	NO
9	26	3,3	Shone syndrome	NO	-1,7	Mitral stenosis, fibro-elastosis LV
10	176	4,5	Aortic stenosis	NO	-1,9	NO
11	15	3,1	Shone syndrome	NO	-3,5	Coarctation
12	206	6,5	Aortic stenosis	NO	-2,1	NO
13	21	3,7	Aortic stenosis	NO	-2,8	NO

Number	Surgery	Complications	Z-score follow-up	Aortic stenosis	Aortic insufficiency
1	Ross-Konno	ECMO, AV-block	1,5	10	1+
2	Ross-Konno	NO	3,98	7	0
3	Ross	NO	1,56	8	0
4	Ross	NO	1,8	12	1+
5	Ross	NO	1,2	5	1+
6	Ross	Renal insufficiency	1,5	6	0
7	Ross-Konno	AV-block	1,4	4	0
8	Ross	NO	2,1	15	1+
9	Ross	NO	0,8	5	1+
10	Ross	NO	1,8	9	1+
11	Ross-Konno	ECMO, Renal insufficiency	0,7	13	0
12	Ross	NO	1,7	7	0
13	Ross-Konno	NO	2,1	5	1+

LHlesions: left heart lesions, LV: left ventricle, ECMO: extracorporeal membrane oxygenation.
Age: days, Weight: kilograms, Aortic stenosis: median gradient (mmHg)

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P2825 - DIGEORGE SYNDROME DOUBLE AORTIC ARCH TOF AND PULMONARY ATRESIA – A CASE REPORT

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Objective: To present a case report of successful management of DiGeorge syndrome with TOF and Double aortic arch.
Methods: A 12 days old male baby born as preterm was diagnosed to have DiGeorge syndrome by amniocentesis and FISH analysis. Echocardiogram revealed TOF with pulmonary atresia. CT pulmonary angio revealed a double aortic arch, forming a complete ring around the trachea and oesophagus with compression. Baby had a pre-op SpO2 of 80%.
Results: After pre-op evaluation, baby underwent division of posterior limb of aortic arch with central shunt and PDA interruption. Post-op recovery was uneventful. Ventilated for 71 hours and later inotropes were tapered and shifted to ward. Post-op SpO2 of 95%.
Conclusion: Antenatal evaluation and prompt neonatal surgical intervention can save these kids with complex cardiac anomaly.

P2826 - D TRANSPOSITION OF THE GREAT ARTERIES SINGLE CENTER EXPERIENCE WITH ARTERIAL SWITCH OPERATION IN THE CURRENT ERA

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The neonatal arterial switch procedure has become the technique of choice for babies with TGA, including transposition with intact ventricular septum, with ventricular septal defect with or without aortic arch hypoplasia, with Taussig-Bing anomaly and with complex coronary artery variants.
Methods: Between 2010 and 2016, 126 consecutive patients who underwent ASO for D-TGA or Taussig-Bing anomaly were included in this study. The median age at the operation was 8 (1-28) days, and 96 patients (75%) underwent ASO less than 2 weeks. The median weight was 3.2 kg (minimal weight 1,5 kg). Eighty two patients with either simple (n = 54) or complex forms (n = 22) of TGA, diagnosed prenatally. Balloon atrial septostomy was performed in 11 cases. The patients were divided into groups: simple TGA (n = 89), complex TGA (n = 31) included those who had TGA with VSD or other anomalies, and Taussig-Bing anomaly with aortic arch obstruction (n = 6). Intramural coronary arteries were detected in 5 of them and single coronary artery in 2 babies. In two cases previously was performed abdominal surgical procedures: resection of Meckel's diverticulum and esophageal plasty. Median follow-up duration was 29 (0.2-72) months.
Results: There were 4 (3,1%) in-hospital deaths - 2 patient with complex coronary anatomy and 2 babies with aortic arch obstruction repair. There was no late deaths. 7 patients (5.7%) required reintervention during follow up: AV insufficiency repair - 1, ASD closure - 1, PA plasty - 1, PA stenting - 1 and PA ballon angioplasty in 3 patients.
Conclusions: Prenatal diagnosis and early neonatal clinical suspicion with adequate measures in the first hours after birth are essential to reduce the early mortality in TGA. ASO can be performed with a low risk of early mortality and satisfactory long-term outcomes. Close long-term surveillance is mandatory to detect hemodynamic changes.

P2829 - HEPATIC VENOUS OXYGEN SATURATION IS LOWER THAN MIXED VENOUS OXYGEN SATURATION IN LATE FONTAN CIRCULATION

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Background: The decrease in hepatic venous oxygen saturation (ShvO2) was proposed to indicate the presence of acute liver dysfunction after cardiac operation especially in Fontan procedure. Hepatic venous oxygen saturation in late Fontan circulation has not been investigated. The purpose of this study is to evaluate the difference between hepatic and mixed venous oxygen saturation in late Fontan circulation.
Materials and Methods: 42 patients with Fontan circulation (F group) and 14 patients with biventricular repair (non-F group) were compared in their results of cardiac catheterization. APC Fontan and fenestrated Fontan, heterotaxy were excluded.
Results: The age at study were 7.2 ± 3.6 years in F group (5.5 ± 2.2 years after the Fontan operation) and 4.9 ± 5.2 years in non-F group. At catheterization, cardiac index was lower for the F group than non-F group (2.9 ± 0.8 ml/min/m2 vs. 3.4 ± 0.7 ml/min/m2; $p = 0.048$) and central venous pressure was higher for the F group (10.3 ± 1.5 mmHg vs. 4.7 ± 1.9; $p < 0.01$). Mixed venous oxygen saturation was lower for F group($65.0 \pm 6.5\%$ vs. $70.1 \pm 4.5\%$; $p = 0.013$) and hepatic venous oxygen saturation was lower for F group($48.8 \pm 14.2\%$ vs. $63.9 \pm 5.9\%$; $p < 0.01$). In F group hepatic venous oxygen saturation was significantly lower than mixed venous oxygen saturation ($p < 0.01$).
Conclusions: Lower cardiac output and higher central venous pressure in Fontan circulation has been interpreted as the cause of lower mixed venous oxygen saturation. In this study, however, we showed the discrepancy between mixed venous oxygen saturation and hepatic venous oxygen saturation. This implies the imbalance of the oxygen supply-demand relation in abdominal organs.

P2833 - INCIDENCE AND FACTORS INFLUENCING SPONTANEOUS CLOSURE OF FENESTRATION AFTER EXTRACARDIAC CONDUIT FONTAN PALLIATION

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Background: Fontan operation is the last stage of single ventricle palliation. Most centers use fenestrated extracardiac Fontan procedure as the preferred modality. Some of these fenestration close spontaneously with time. There is limited data available about the incidence of spontaneous closure of this fenestration and the factors which influence this. The aim of this study is to identify the incidence of spontaneous closure of Fontan fenestration and the factors influencing it.
Methods: Retrospective review of patients who underwent fenestrated Fontan surgery at our center between January 1, 2001 and June 30, 2016 was done. Data extracted include age at Fontan, preoperative data including mean pulmonary artery pressure, transpulmonary gradient, pulmonary vascular resistance, ventricular end-diastolic pressure, Nakata and McGoon Pulmonary artery indices, presence of significant aortopulmonary collaterals. Perioperative and post-operative variables obtained include cardiopulmonary bypass, cross clamp time, pleural effusion lasting

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>2weeks, ventricular function and atrioventricular valve regurgitation, thromboembolism. The last clinic visit was reviewed for NYHA class, arrhythmia history, oxygen saturation and evidence of protein losing enteropathy. The most recent echocardiogram/or MRI was reviewed for any evidence of Fontan pathway obstruction and patency of fenestration.
Results: Of 82 subjects who underwent fenestrated Fontan surgery, 15 did not have postoperative follow-up at our center. Of the remaining 67 patients, there was no echocardiographic evidence of patent fenestration in 27 patients (40%). Of these 27 patients, 16 (59%) had spontaneous closure of the fenestration and 10 patients (37%) underwent closure of fenestration in the cardiac catheterization laboratory and 1 (4%) had surgical closure of the fenestration. None of the variables evaluated had significant predictive value for spontaneous closure of the fenestration.
Conclusions: 24% of patients had spontaneous closure of fenestration after Fontan palliation. There was no variable in these patients which could have predicted the potential for spontaneous closure.

P2847 - EARLY EXPERIENCE USING COMPOSITE VALVED RV TO PA CONDUIT IN STAGE I RECONSTRUCTION

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Background: Use of a valved right ventricle to pulmonary artery (RV-PA) conduit as part of the Stage I palliation (S1P) for univentricular heart heart disease may diminish ventricular volume loading, improve pulmonary blood flow, or lower mortality compared to a Sano shunt.
Methods: Infants with univentricular heart disease undergoing S1P using either a composite, valved RV-PA conduit (proximal ringed PTFE graft and distal cryopreserved femoral vein graft, n = 18) or standard Sano conduit (control, n = 53) at Boston Children's Hospital between 2013 and 2016. Endpoints included markers of oxygen delivery assessed by linear mixed model analysis accounting for repeated measures over time from postoperative day 0-7.
Results: On average from POD 0-7, the valved Sano group had a higher arterial (by 4.5%, P = 0.02) and venous oxyhemoglobin saturation (by 5.2%, P = 0.12), and a similar serum lactic acid (lower by 0.9, P = 0.24) than did standard Sano group. The time to cardiac catheterization was similar between groups (P = 0.38, log rank test). Of the valved Sano group, 5 patients required stenting of conduit (two at RV connection and three at femoral vein to PA anastomosis). There was evidence of valvar patency in all of the valved Sano group by echocardiography at the time of evaluation for the Glenn operation. Survival to hospital discharge was similar between groups (78.6% vs 88.9%, P = 0.63).
Conclusions: The use of a valved Sano conduit as part of S1P is safe and may improve acute postoperative hemodynamics. Further modifications to the distal anastomosis may diminish the need for stenting.

P2848 - FATE OF 775 MAPCAS IN 271 PATIENTS THE INTER RELATIONSHIP OF NATIVE PULMONARY ARTERIES WITH COLLATERAL VESSELS IN PULMONARY ATRESIA MAPCAS

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Background: The relationship between native vessels and MAPCA supply is very variable. We utilised an individualised approach to maximise recruitment of the vascular bed, utilising both native PAs and MAPCAs and reviewed 30 year clinical outcomes and MAPCA patency/re-intervention.
Methods: Between 1988–2016, 271 patients with PA/MAPCAs were treated. The strategy aimed to achieve unifocalisation using native PAs if present and recruiting all MAPCAs where there was no dual supply. The VSD was left open if <15/20 lung segments recruited. Unifocalisation was categorised in 3 groups: (Group1) based on native system only, (Group2) combination of native system and MAPCAs; and (Group3) MAPCAs only (absent native PAs).
Results: Unifocalisation was achieved in 92% of patients with 30-day mortality of 2.8% and 1,5,10 and 15 year survival of 97.1, 95.69, 93.0 and 91.3% respectively. A total of 474 MAPCAs (29% Group1, 48% Group2 and 23% Group3) were recruited with the remainder ligated due to dual supply. In 22(8%) patients, unifocalisation was not achievable and they were palliated with shunt procedures, although 6(27%) now await unifocalisation. It was possible to close the VSD in similar numbers across the 3 groups: 81.9%, 78.1%, 79.3%, with survival at 10 years of 98.6%, 93.7%, 84.8% respectively(0.0607 log-rank test). There was no 'era effect' on survival. DiGeorge Syndrome was strongly associated with failure to close VSD(p = 0.02). Of 97 patients where the VSD was initially left open, 46(47.4%) underwent subsequent closure. Overall, VSD closure was achieved in 198 (79.5%) of patients, incorporating 364 MAPCAs.
Conclusion: Although native PAs provide the best substrate for unifocalisation, recruitment of MAPCAs is essential to achieve successful VSD closure. In a small proportion (6%) with intrinsically poor vasculature, unifocalisation cannot be achieved. A strategy of initial unifocalisation, leaving VSD open can be helpful in borderline cases with successful septation of the circulation being achieved in the majority.

Table.

Groups				
Unifocalised	**All patients (n = 249)**	**Group 1 (n = 72)**	**Group 2 (n = 119)**	**Group 3 (n = 58)**
DiGeorge	64 (25.7%)	15 (23.4%)	30 (46.9%)	19 (29.7%)
MAPCAs sum/mean	775 (3.11)	177 (2.45)	404 (3.39)	194 (3.34)
Shunt: mBT	73 (29.3%)	17 (23.3%)	36 (49.3%)	20 (27.4%)
Shunt: Melbourne	56 (22.5%)	33 (58.9%)	23 (41.1%)	0 (0.0%)
No shunt	120 (48.2%)	22 (18.3%)	60 (50.0%)	38 (31.7%)
VSD status (unifocalised)				
VSD primary closure	152 (61.0%)	52 (34.2%)	67 (44.1%)	33 (21.7%)
VSD closed: delayed closure	46 (18.5%)	7 (15.2%)	26 (56.5%)	13 (28.3%)
VSD remains open	51 (20.5%)	13 (25.5%)	26 (51.0%)	12 (23.5%)
Group 1	PA/VSD/MAPCAs : Native PA system only (no recruited MAPCAs)			
Group 2	PA/VSD/MAPCAs : Combination of native PA system and MAPCAs			
Group 3	PA/VSD/MAPCAs : MAPCAs only (absent native PAs)			

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P2851 - TRICULSPID VALVE DETACHEMENT DURING CLOSURE OF VENTRICULAR SEPTAL DEFECTS VALVULAR COMPETENCE AT LONG TERM FOLLOW UP

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Background/Hypothesis: Tricuspid valve detachment (TVD) has been utilized during ventricular septal defect (VSD) repair for improved visualization of and access to the defect. The objective of this study was to determine if TVD was associated with increased tricuspid regurgitation at long-term follow-up.
Materials and Methods: 272 patients (age <18 years) undergoing perimembranous VSD repair at our Institution between 1995-2013 were identified. Eighty-six patients (31.6%) underwent concomitant TVD to enhance exposure. The degree of tricuspid valve regurgitation at latest echocardiographic follow-up (trivial, mild, moderate, and severe according to the 2003 Journal of the American Society of Echocardiography recommendations) was reviewed. Intergroup differences were evaluated with the Fisher exact test or the Mann-Whitney U test for categorical and continuous variables, respectively.
Results: Fifty-seven percent of subjects were male, with a mean age at intervention of 743 + 1101 days and no significant difference between TVD and non-TVD groups. Mean echocardiography follow-up was 7.5 + 5.2 years. There was no significant difference in degree of tricuspid regurgitation in subjects with and without TVD with the mean degree of tricuspid regurgitation of both groups falling within the trivial to mild range ($p = 0.54$). Furthermore, there was no significant difference in the incidence of residual VSD between the two groups ($p = 0.64$).
Conclusions: Tricuspid valve detachment is a valid technical adjunct to VSD closure and is associated with excellent long-term valvar competency and no increase in the incidence of residual VSD. Surgeons should not hesitate to detach the tricuspid valve when exposure of the defect is limited.

P2878 - LEFT ATRIAL ISOMERISM A LONG TERM OUTCOME STUDY

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Left atrial isomerism (LAI) is a rare disorder associated with a wide variety of cardiac and extracardiac abnormalities with varying outcomes.
Objective: The aim of the study was to describe the cardiac and non-cardiac associations in patients with LAI. We also aimed to determine the long term outcomes and analyse the risk factors associated with adverse outcome.
Methods: Retrospective casenote review of cases with LAI managed at Birmingham Children's Hospital from 1980-2015. The outcome measures were overall survival and survival following surgical intervention.
Results: Over a 35 year period, 94 patients were identified with LAI. The cardiac abnormalities are described in table 1. Commonest extracardiac abnormalities were polysplenia in 57(61%) and biliary atresia in 11(12%). 48 (51%) had biventricular circulation (6 were not operated) while 46 (49%) were palliated along the univentricular route. Overall mortality was 30 (32%), with 13 (27%) in biventricular circulation and 17 (37%) in those with univentricular circulation. Overall survival following surgical intervention was 83% at 1year, 75% at 5yrs and 72% at 10yrs. There was no difference in the 1 and 5 year survival in both the groups, however the 10 year survival was 68% in the univentricular group and 75% in the biventricular group. Risk factors for adverse outcome were congenital complete heart block ($p = 0.03$), total anomalous pulmonary venous drainage ($p = 0.01$) and need for neonatal cardiac surgery ($p = 0.04$). Of those who underwent surgery, there was no statistically significant difference in the survival between the single and biventricular circulation groups ($p = 0.44$).
Conclusions: There is no difference in the medium term outcomes of the LAI, irrespective of univentricular or biventricular circulation. Survival in both groups is influenced by the presence of anomalous pulmonary venous drainage, complete heart block and need for neonatal cardiac surgery.

Table.

	Number (percentage)		Number (percentage)
Systemic veins	**Pulmonary artery**		
Bilateral SVC	51 (53)	Pulmonary atresia	11 (11)
Left SVC	9 (9)	Pulmonary stenosis	29 (31)
Right SVC	33 (33)		
Interrupted IVC	82 (85)		
Pulmonary veins	**Ventriculoarterial (VA) Junction**		
PAPVD	14 (14)	Double outlet right ventricle	23 (24)
TAPVD	8 (8)	VA discordance	18 (18)
Atrial septum	**Left ventricular outflow**		
Atrial septal defect (Isolated)	19 (20)	Hypoplastic arch	17 (17)
Common atrium	41 (42)	Subaortic stenosis	5 (5)
Atrioventricular (AV) junction	**Isolated VSD**	10 (11)	
Complete atrioventricular septal defects (AVSD)	47 (48)		
Partial AVSD	16 (16)		

P2883 - RATIONALE AND DESIGN OF A STEM CELL TRIAL IN HYPOPLASTIC LEFT HEART SYNDROME PATIENTS

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Background: The survival rate of hypoplastic left heart syndrome (HLHS) patients remains limited. We describe the rationale from a preclinical model and study design for the first Phase I trial in the United States to evaluate the safety and feasibility of allogeneic mesenchymal stem cell (MSC) therapy in HLHS patients.
Materials and Methods: Neonatal Yorkshire swine (n = 20, 6-9 kg) underwent pulmonary artery banding to induce RV dysfunction. Human stem cells were injected intramyocardially into the RV at

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30 minutes post-banding (MSCs: n = 5, cardiac stem cells (CSCs): n = 5; MSCs/CSCs: n = 5; placebo: n = 5). RV size and function was monitored with serial transthoracic echocardiography.
Results: Four weeks post-injection, stem cell-treated myocardium had a smaller increase in RV end-diastolic area, end-systolic area, and tricuspid vena contracta width ($p < 0.01$), increased RV fractional area of change ($p < 0.01$), and improved myocardial strain mechanics ($p < 0.01$) relative to placebo. Myocardium from MSC-treated piglets exhibited several mechanisms of endogenous myocardial repair, including enhanced neovessel formation, recruitment of endogenous c-kit$^+$ CSCs and proliferation of cardiomyocytes and endothelial cells. Mechanistically, MSC treatment of porcine RV myocardium as well as an in vitro model of cardiomyocyte hypertrophy was associated with upregulation of the cardioprotective, anti-hypertrophic molecule growth differentiation factor 15. Based on these preclinical results, an FDA approved trial has been initiated in a total of 30 HLHS patients who will be randomized to receive intramyocardial allogeneic MSCs at the Stage II palliative bidirectional cavopulmonary connection (BDCPC) operation versus control. This study will examine safety and feasibility endpoints as well as preliminary efficacy to enhance RV performance. To date, three infants (n = 3) with HLHS have safely undergone allogeneic MSC injection at the time of the Stage II BDCPC operation.
Conclusions: This Phase I trial will provide important insights of allogeneic MSC therapy in HLHS patients. The lessons learned may be applied to other congenital heart diseases.

P2891 - LONG TERM FOLLOW UP OF BIDIRECTIONAL CAVOPULMONARY ANASTOMOSIS PATIENTS MULTI CENTER STUDY

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Background: The bidirectional cavopulmonary shunt is a step towards the Fontan operation in palliation of patients with single-ventricle heart.In Egypt, there is a delayed age of children undergoing Fontan procedure due to long waiting lists and budget restraints on pediatric cardiac surgeries.
Aim of the work: To assess long term follow up of patients with bidirectional cavopulmonary shunt and to determine their clinical and hemodynamic status.
Methods: A total of 125 patients with bidirectional cavopulmonary shunts from three centers in Egypt were followed up from January 2012 till July 2016. At follow up, mean age was 7.5 (1.7-16 years) and weight was 23.05 (9-58 Kg). Males were 61.5%, 64% had single ventricle morphology, 76% had additional pulmonary flow and 38.5% had prior palliative cardiac surgery.
Results: The mean age at Cavopulmonary shunt was 2.98 years (0.4–12 years). NYHA was class I in 82% and II in 15%, mean oxygen saturation was 81% (60-95). Cardiac catheterization was done in 45%, mean Glenn pressure was 15.5(6-28 mmHg) and mean Nakata index was 287 (108-910). Catheter interventions were done in 24%; eleven patients had procedures to minimize the extrapulmonary flow: closure of the shunts, collaterals, patent ductus arteriosuus and/or device occlusion of the forward flow, while thirteen patients had balloon and/or stent of pulmonary arteries, superior vena cava (SVC) or pulmonary veins. five patients had occlusion of venoatrial collaterals and/or left SVC to coronary sinus. The only significant difference was intensive care unit (ICU) stay which was shorter in patients with additional pulmonary flow (P value 0.016), and mean Glenn pressure which was lower in patients with single ventricle (P value less than 0.001). Mean follow up was 13.33 years with 5% late mortality.
Conclusion: Cavopulmonary shunt can be a long term palliative procedure for patients with single ventricle in developing countries..

Figure 1.

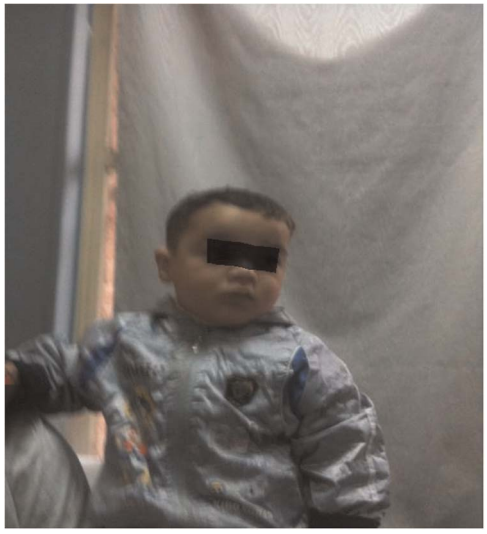

Figure 2.

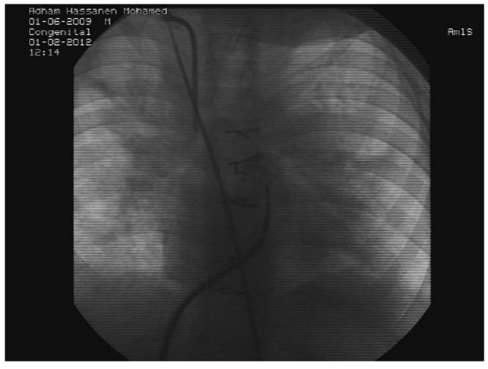

Figure 3.

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P2903 - COMPREHENSIVE INTERSTAGE CARDIAC CARE PROGRAM FROM FETUS TO GLENN RESULTS IN IMPROVED SURVIVAL FOR COMPLEX SINGLE VENTRICLE INFANTS

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Background: Interstage mortality for single ventricle (SV) patients is 10-20%. Care protocols have been reported to improve outcomes in this population.
Methods: In December 2012, a change in surgical strategy along with an interstage monitoring protocol following the Norwood operation was created in conjunction with our SV clinic. Guidelines from prenatal to stage-II palliation included a home surveillance program, trigger points for urgent office visits and admissions via a SV hotline, as well as frequent echocardiograms and nutrition evaluations. Outcomes for infants undergoing stage-I palliation before and after initiation of the care guidelines were reviewed. End points included operative mortality, postoperative length of stay, interstage discharge to home, interstage mortality, growth, and timing of stage II palliation.
Results: Forty-one infants between January 2008 and December 2012 comprised the pre-SV program cohort. Since the SV program, 54 patients have undergone stage I palliation with a significant reduction in operative mortality (16% vs. 5.7%). Notably, 26 Norwood procedures have been performed since September 2014 with no operative mortality (STS national benchmark 15.6%). Postoperative median length of stay in the post-SV program cohort is 26 days with 87% of patients discharged home. Interstage mortality decreased from 26% to 4% post-SV program. The last interstage mortality was in July 2014 with 28 Norwood operations performed since then. Time to bidirectional Glenn was shortened from 143 days (range 99-261 days) to 130.5 days (range 89-185 days) at a comparable patient weight. Furthermore, there was a significant reduction in growth failure (25% to 4.3%).
Conclusion: Outcomes have significantly improved at our institution for stage I palliation after changing surgical strategy and initiating a comprehensive care plan (inter-stage monitoring protocol and SV clinic). Although many centers already have interstage programs, this comprehensive fetus to Glenn program has improved the.

P2906 - PULMONARY AUTOGRAFT MITRAL VALVE REPLACEMENT (ROSS II) INTERMEDIATE AND LONG TERM FOLLOW UP OF A U.S. CENTER

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Background: Mitral valve replacement (MVR) in pediatric and adolescent patients poses significant challenges, specifically regarding valve size, durability, and need for anticoagulation. The use of a pulmonary autograft or allograft for mitral replacement, the Ross II MVR, has been reported by centers seeking durable replacements without requiring anticoagulation. We report medium- to long-term follow-up in the largest reported experience with this procedure in North America.
Methods: We conducted a retrospective review of Ross II patients from January, 2003 to January, 2010. Autografts and allografts were constructed by suturing valves inside a 2.5 cm Dacron tube and covering the outside with pericardium. The composite prosthesis was sutured to the mitral annulus with interrupted sutures.
Results: Mean age at the time of surgery was 25.15 ± 15.72 years. 80% of patients were female. Mean follow-up was 11.74 ± 5.2 years. There were no in-hospital deaths. There were 3 late deaths post Ross II procedure, occurring at 11 months (respiratory failure), 5 years (unknown cause), and 11 years (cardiac arrest), respectively. Three patients required subsequent mechanical MVR at a median interval of 2 years after the Ross II procedure. Echocardiographic follow up was available for 5 of 7 patients who retained the Ross II prosthesis at a mean of 10.53 ± 3 2 years postoperatively and revealed mean mitral valve gradients ranging from 2.2 to 9.6 mmHg. One patient had greater than mild mitral regurgitation; all others had minimal-to-no mitral regurgitation.
Conclusions: The Ross II procedure may be a valid option for a select subset of older pediatric and adolescent patients for whom a durable tissue valve devoid of the bulk or need for anticoagulation is necessary or desired. Our experience indicates this technique may offer a durable solution for these challenging patients.

Table 1. Demographic and Echocardiographic Information.

Patient	Age at last follow up (years)	Age at Ross II (yrs)	Follow Up (yrs)	Echocardiopraphic Follow-up (years)	Mean Mitral Gradient (mm Hg)	Mitral Function
1^	60.91	46	14.91	–	–	–
2	56.83	43	13.83	12.66	2.2	Normal
3	21.66	8	13.66	11	9	Moderate stenosis
4^	28.75	14	13.75	–	–	–
5	34.66	19	15.66	13	6.5	Mild stenosis
6*	27.25	16	11.25	11	9.6	Mild-moderate regurgitation
7^	33.75	15	18.75	–	–	–
8*	1.75	0.58	1.17	–	–	–
9*	46.53	41	5.53	5		Minimal regurgitation
10	37.91	29	8.91	–	–	–

*Deceased.
^Patient underwent subsequent mechanical mitral valve replacement.

P2919 - HYPOPLASTIC AORTIC ARCH REPAIR BY THORACOTOMY IS IT PLAUSIBLE

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Objective: The aim of this study is to present the early clinical outcomes of patients with severe aortic coarctation and hypoplastic aortic arch that underwent repair through a thoracotomy.
Methods: A retrospective cohort of patients with aortic coarctation or coartation and hypoplastic aortic arch with complete echocardiographic evaluation of the aorta, who underwent repair by thoracotomy between 2009 and 2016 was analyzed. Pre-operative, post-operative, and echocardiographic characteristics were analyzed. Mortality and reintervention outcomes were evaluated by phone at one year follow up. Hypoplastic aortic arch was defined as distal transverse arch/ascending aorta ratio ≤ 0.6 and was compared to simple coarctation cases.

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Results: In the study period 380 patients underwent combined or isolated aortic coarctation repair. For the analysis 55 patients meet the inclusion criteria. 40% (22/55) were newborn, 38.2% (21/55) infants and 21.8% (12/55) older than 1 year. 7.2% (4/55) had chromosomal anomalies. Preoperative ejection fraction was 68.6 ± 10.6%. Mean distal transverse arch/ascending aorta ratio was 0.6 ± 0.2. There was no global mortality or follow up 1 year mortality. Reoperation rate was 3.6% (2/55). 30.6% were termino-terminal repairs, 4.1% were subclavian flap repairs, 63.3% were extended termino-terminal repairs and 2% were extended termino-terminar repairs with subclavian flap. Re-coarctation rate for termino-terminal repair was 5.9% and for extended termino-terminal repair was 3.1% without any statistical difference.
Conclusion: Patients undergoing hypoplastic aortic arch repair through thoracotomy had similar outcomes when compared to patients undergoing isolated aortic coarctation repair. Taking into account the morbidity of a median sternotomy and extracorporeal pump needed to repair a hypoplastic arch through a sternotomy it is reasonable to use a thoracotomy for repair. Even though the proportion of patients with re-coarctation was higher with a termino-terminal approach, there was no statistical difference but extended termino-terminal is recommended.

Table.

	Distal transverse arch/ ascending aorta ratio	
Characteristic	**<0.6**	**>0.6**
Patients n (%)	14 (25.4)	41 (74.5)
Age in months Median	1.2 (0.4-3.9)	1.9 (0.7-7.4)
Female n (%)	8 (57.1)	13 (31.7)
Distal transverse arch Median mm	3.6 (3.4-6.2)	6 (5-7.6)
Isthmus/ascending aorta ratio Median	0.4 (0.3-0.44)	0.48 (0.37-0.64)
In-hospital stay Median	8 (6-25)	8(6-13)

P2920 - A SURVEY OF CURRENT PRACTICES FOR PRE BIDIRECTIONAL GLENN EVALUATION OF SINGLE VENTRICLE PATIENTS IN NORTH AMERICA

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Background: Cardiac magnetic resonance (CMR) is reported to be a less costly, effective alternative to cardiac catheterization (cath) in selected single-ventricle patients prior to bidirectional Glenn (BDG). To what extent CMR is used in routine pre-BDG evaluation at cardiac centres in North America is unknown.
Materials/Methods: We conducted an online survey from July to November 2016, exploring North American practices for pre-BDG evaluation among pediatric cardiologists and cardiovascular surgeons treating children with congenital heart disease.
Results: In total, 184(19.6%) practitioners responded (cardiologists: 89%, surgeons: 11%). Of these, 87% reported ≥10 years of practice and the majority were from centres performing ≥100 open-heart surgeries(88.1%) and ≥10 BDG(86%) operations annually. Sixty percent reported existance of an institutional policy for pre-BDG evaluation including: echocardiography with cath for all patients (76.4%), echocardiography with CMR for low-risk patients (14.5%), echocardiography with CMR for all patients (1%), and other modalities (8.1%). Individually, With 69.3% and 12.5% of practictioners, respectively recommended cath or CMR in >75% of instances. If any risk factor (aortopulmonary or venovenous collaterals, pulmonary vein stenosis, pulmonary hypertension, arch obstruction, ventricular dysfunction or restrictive atrial communication) was identified by pre-BDG echocardiography, 72% would refer for cath alone, 26% would refer for cath with CMR, and 2% would for other modalities. 96% and 99% estimated the frequency of a preoperatively missed diagnosis as <25% of instances with CMR or cath, respectively. The minority (17.3%) of practitioners felt that, in most cases, early unplanned re-operation following BDG resulted from missed diagnoses due to inadequacies of the pre-BDG imaging while 56.3% disagreed and 26.4% provided a neutral response.
Conclusions: There's practice variations across North America for pre-BDG evaluation of single-ventricle patients. Echocardiography combined with cath is most commonly used in spite of existing data to support use of CMR for at least low risk.

P2927 - AORTOPULMONARY WINDOW IN AN ADULT PATIENT. IS SURGERY POSSIBLE AT THIS STAGE OF LIFE

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Introduction: In concept, an aortopulmonary window is a communication between the main pulmonary artery and the ascending aorta in the presence of two semilunar valves separately. There are no large series collected specifically for adults with this pathology because of their fatal nature within the first year of life, but surgical correction has been described and feasible, with certain particular considerations and with good results in the short, medium and long term.
Case: A female patient of 22 years of age presents with flat dyspnea and palpitations sensation, cyanosis labial to the effort. She is originally from a rural sector of the capital. A radiographic study determined cardiomegaly GIII with biventricular growth and increased pulmonary vasculature. It is diagnosed by Transthoracic Echocardiography and Cardiac Catheterization of a congenital Aortopulmonary Window type II, with systolic pressure of the pulmonary artery between 80 and 90 mmHg and a short circuit from left to right, QP/QS 2.87, RPT 634.3 dynes, 7.9 uw, RST 1960. The only treatment option was surgical. The finding of the surgery was a aortopulmonary window type III. The total time used for the surgical correction was 197 min, with a cardiopulmonary bypass time of 96 minutes, aortic clamping of 62 minutes and reperfusion in 18 minutes, working with hypothermia up to 32 °C. She remains hospitalized in intensive care unit for 24 hours. Was discharged within 4 days of leaving the unit. The final echocardiogram at the end reports a pulmonary artery pressure of 36 mmHg, without short circuits, without aortic and pulmonary insufficiency. Currently in a postoperative period of 12 months, with good general condition, without cyanosis, she does his daily activities without problem and does not require oxygen. The patient lives in a rural parish that is 2830 meters above sea level.

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P2939 - LONG TERM SURVIVAL BY PHYSIOLOGIC CLASSIFICATION AFTER SURGICAL OR TRANSCATHETER INTERVENTIONS FOR CONGENITAL HEART DISEASE (CHD) A STUDY FROM THE PEDIATRIC CARDIAC CARE CONSORTIUM (PCCC)

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University of Minnesota, Clinical and Translational Science Institute, Minneapolis-United States[2]; University of Rochester, Pediatrics, Rochester-United States[3]; Children, Surgery, Kansas City-United States[4]; University of Minnesota, Pediatrics, Minneapolis-United States[5]; University of Minnesota, Internal Medicine, Minneapolis-United States[6];

Background: We examined the long term survival of patients surviving interventions for CHD as reported in the PCCC between 1982 and 2003. The PCCC is the longest standing registry of outcomes for pediatric cardiac interventions in the world containing data from surgical and trans-catheter procedures from 48 centers in 27 U.S. states.
Methods: We linked the PCCC dataset with the National Death Index and the United Network for Organ Sharing using direct identifiers to evaluate long term vital status of individuals with repaired or palliated CHD. A total of 56,834 subjects were registered prior to April 15, 2003 and matched through December 31, 2014. Time from birth to first transplant or death was compared using Cox Proportional Hazards Regression and Kaplan-Meier survival plots to compare major physiologic classifications. Follow-up time was administratively censored after 50 years.
Results: A total of 9,049 deaths or transplants (15.9%) were identified within 50 years of birth. Time to event or censoring ranged from 1 day to 50 years with a median time of 21.1 years. Analysis by physiologic group revealed a hazard ratio of 0.167 (95% CI: 0.160-0.175) for two-ventricular conditions compared to single-ventricle lesions. Within the group with two ventricle physiology, hazard ratios relative to complete mixing were: 0.30 for impaired pulmonary flow (95% CI: 0.27-0.33), 0.53 for impaired pulmonary venous return (95% CI: 0.44-0.64), 0.21 for impaired systemic flow (95% CI: 0.19-0.25), 0.14 for pulmonary left-to-right shunt physiology (95% CI: 0.12-0.16), and 0.50 for TGA physiology (95% CI: 0.43-0.57).
Conclusions: Mortality is higher for patients with single ventricle physiology. Within patients with 2-ventricle physiology mortality differs significantly based on the underlying main physiology with the highest mortality observed in the group of complete mixing and the lowest for left-to-right shunt physiology..

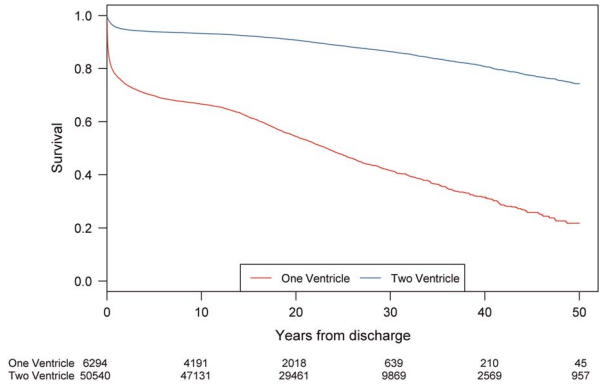

Figure.

P2942 - CORONARY ARTERY PATTERN IN TRANSPOSITION OF THE GRAT ARTERIES AND ITS INFLUENCE IN ARTERIAL SWITCH

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Clinical background: Now on days arterial switch is the election treatment for surgical correction of transposition of the great arteries. For the success of anatomical correction of transposition of the great arteries it is essential to transfer the coronary ostia to the posterior vessel without undue tension, torsion, or kinking of the proximal coronary arteries or their early branches, requiring understanding the anatomical disposition and variations of the coronary arteries in the cited disease.
Objetive: Describing the origin and the anatomical variations of the coronary arteries in patients with transposition of the great arteries, and its relation with the mortality in anatomical correction.
Material and Methods: Retrospective and descriptive study. The anatomy of the coronary arteries was examined and described during operation of arterial switch, between January 2007 and December 2016, in 155 patients (99 male/56 female), with a mean age of 8 months. (1 day – 7 years).
Results: Coronary artery nomenclature was based on the Leiden Convention Classification. The usual coronary distribution and the most common variant (1LAD,CX; 2RCA) was found in 77.4% of the patients. Eight patients (5.2%) had a 1LAD,CD; 2CX pattern, six patients (3.9%) had 1LAD; 2CX,RCA; a pattern 1LAD,CX,RCA in a unique ostium in 4 subjects(2.6%), and a 2LAD,CX,RCA in a unique ostium in four patients; other patterns were less frequent. During the correlations of the pattern and mortality of the subjects, no statistic significance was found. (p 0.89).
Conclusion: The anatomic variants frequency was similar to the reported in literature. A great diversity of isolated patterns with a complex origin and trajectory was found; however, no relationship between mortality and coronary pattern was observed.

P2945 - CHARACTERISTICS AND EVOLUTION OF THE SURGICAL MANAGMENT FOR THE TRANSPOSITION OF THE GREAT ARTERIES

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Transposition of great arteries is one of the most common cardiovascular congenital malformations, with an incidence of 1 in 3500-5000 newborns. In the last three decades, a survival increase has been reported in this patients, due to an earlier diagnosis, prostaglandine e1 administration on a regular basis, and a strong progress in the technical aspects of the cardio surgical units and cardiac catheterization.
Objective: describe the background and evolution of affected subjects who were through surgery, and determine possible prognostic factors in morbility and mortality in a short and midterm.
Material and Methods: retrospective and descriptive study. Review of clinical files of subjects with great arteries transposition who were operated between 2007 and 2016.

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Results: 192 subjects diagnosed with great arteries transposition were selected (105 IVS/85 VSD). In 127 of the patients, a surgical correction during the first surgical procedure was achieved (anatomical 91%/physiological 9%), with a mortality of 22.8%. In 24 patients, the anatomical correction was done after ventricular preparation (mortality of 33.3%). A higher mortality is observed in patients with transposition with aggregates malformations, mainly coarctation of the aorta. In the linear regression analysis, we found significant significance for mortality in the following variables: cardiopulmonary bypass time, aortic cross clamp time, postoperative mechanical ventilation time, and the age of surgery.
Conclusion: In our institution, the mortality in the surgical management of the transposition of the great arteries is higher than the reported in the literature; this is closely related to the learning curve and improvement of the surgical technique over the years.

P2954 - TOTAL RIGHT VENTRICULAR EXCLUSION FOR END STAGE ARVD AND EBSTEIN ANOMALY

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Introduction: The best maagement for end stage RV dysfunction is under debate. This condition can be seen as Arrhythmogenic RV dysplasia or in extreme cases of Ebstein anomally.Total RV exclusion has been introduced as a final management modality in these patients. We report our experience in 2 patients.
Method and Result: A 13 y/o girl who presented with severe dyspnea and CHF and a 2 year old girl with extreme form of non repairable Ebstein anomaly are reported. Both patients had very dilated dysfunctional right venricles pushing the interventricular septum towards left. Both had right to left shunts through ASDs causing relative cyanosis.The first patient was candidate for heart transplant and had frquent admissions due to severe sympyoms. Echo, Angiography & MRI confirmed the diagnosis in both patients. The diagnosis of ARVD(arrhythmogenic right ventricular dysplasia) was made in Patient 1 although full criteria could not be met. The whole RV was excised and the tricuspid valve closed by fenestrated patch and Extra-cardiac ferestrated TCPC performed at the same stage in both patients. A patch used to replace the RV wall in patient 1 and direct closure done inpatient 2.Fortunately they tolerated the operation well and discharged on the 12th and 14th post op days.
Conclusion: Total right ventricular exclusion can be done with satisfactory result in patients with exceptionally non-functional RV disease.

P2955 - INITIAL EXPERIENCE IN THE SURGICAL MANAGEMENT OF CONGENITAL HEART DISEASE IN NORTHWESTERN MEXICO

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Objectives: To evaluate the 5-year results of the beginning of activities of a surgical center for congenital heart disease in Northwestern Mexico.
Methods: We collected data from all patients who underwent congenital heart disease correction surgery from January 2012 to December 2016, classifying them by age, weight, complexity of heart disease and risk of mortality RACHS-1 and Aristoteles.
Results: During 5 years of operation, 165 surgical procedures were performed. The age range ranged from 9 days to 14 years, with a mean age of 3 years. (57%) were male. Regarding the complexity of heart disease (17%) Complex heart disease, (51%) heart disease of moderate complexity and the rest (32%) Simple heart disease. Mortality associated with procedural complexity and stratified risk corresponds to the international average.
Conclusions: It is possible to decentralize the surgical management of congenital heart disease in Mexico, adequately addressing cases of simple and moderate complexity with similar results to the international average. Complex cases still have to be treated in national reference centers.

P2956 - SURGICAL REPAIR OF THE COMMON ARTERIAL TRUNK IN THE NATIONAL INSTITUTE OF CARDIOLOGY OF MEXICO

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Objective: To describe the anatomical characteristics, surgical treatment and evolution of patients who underwent surgical correction by diagnosis of Common Arterial Trunk at the National Institute of Cardiology Ignacio Chávez, from Mexico City from January 2003 to December 2016.
Material and Methods: A descriptive, longitudinal and retrospective study was carried out. The study population consisted of all patients of the National Institute of Cardiology Ignacio Chavez, who underwent surgical correction by Diagnosis of Common Arterial Trunk from January 2003 to December 2016.
Results: Of the total of the sample, 59 patients were submitted to surgical correction by Diagnosis of Common Arterial Trunk. Of these, 31 were females and 28 males, the mean age was 8 months with a maximum age of 15 months and a minimum of 1 month, the average weight was 5.6 kg, with a maximum of 10 kg and one Minimum of 2.5 kg, the type of correction was with Rastelli technique in 86% of cases and with Barbero Marcial technique in the remaining 14%, the average diameter of the Rastelli tube was 10.85 mm in diameter, and the material used Was Dacron in 60.8%, Goretex in 16.3% and 22.9 xenograft, 4.5% of patients required interventionism in the immediate postoperative period due to a significant gradient of obstruction in the Rastelli tube or in the pulmonary branches. The need for reoperation was present in 22% of the patients during the follow-up to change the Rastelli tube.

P2963 - SURGICAL STRATEGY FOR PA VSD MAPCA WITH ABSENT OR HYPOPLASTIC CENTRAL PULMONARY ARTERY

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Objectives: Pulmonary atresia and ventricular septal defect with major aortopulmonary collaterals (PA/VSD/MAPCA) is a

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complex lesion that is challenging to manage surgically, especially when the central pulmonary artery (cPA) is absent or hypoplastic. We report our experience with one-stage unifocalization of the MAPCA, cPA plasty and palliative right ventricular outflow tract reconstruction (pRVOTR), followed by definitive repair at a later stage.
Methods: From 2000 through 2016, the cPA was reconstructed under cardiopulmonary bypass in 26 patients (mean age 1.2 years, mean weight 8.8 kg) using a Y-shaped autologous pericardial roll. MAPCAs were anastomosed directly to the roll by pRVOTR, resulting in an RVOT diameter 75% that of the normal pulmonary annulus. MAPCA banding was proceded in one patient and Blalock-Taussig shunts in five. The strategy for unifocalization of the MAPCA and pulmonary reconstruction was planned preoperatively using 3D-CT angiographic images.
Results: The mean follow-up period was 6.9 years. There was one surgical death and one late death. Nineteen patients achieved definitive repair with complete VSD closure after 1.3 ± 0.4 years, and 3 patients were waiting. No patient required reoperation. The PA systolic pressure was 27.5 ± 11.4 mm Hg after unifocalization + pRVOTR, decreasing to 23.9 ± 5.6 mm Hg after definitive repair. Oxygen saturation was 79% before and 88% after unifocalization + pRVOTR.
Conclusions: Staged surgical treatment of PA/VSD/MAPCA with absent or hypoplastic cPA resulted in excellent early to mid-term results. pRVOTR might be a good option to obtain adequate pulmonary blood flow.

P2969 - LATE RESULTS AND 4D FLOW MRI EVALUATION OF DOUBLE DECKER TECHNIQUE FOR PARTIAL ANOMALOUS PULMONARY VENOUS RETURN TO THE SUPERIOR VENA CAVA

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Objective: Conventional procedures for PAPVR to the SVC still have problems such as late SVC obstruction, supraventricular arrhythmia, and PVO. We developed a new procedure with minimum right atriotomy and double-barreled arrangement of both systemic and pulmonary venous channels (double-decker technique).
Methods: Between November 1998 and October 2016, 18 patients underwent this operation. The median age was 4.0years, and body weight was 17.5 kg. The right atrial wall was incised from the appendage to few millimeters short of the SVC-atrial junction. The external upper wall of the proximal SVC was covered with the appendage, taking care to avoid injury to the sinus node and sinus nodal artery. New SVC channel was reconstructed onto the proximal SVC external wall. The atrial septal flap or autologous pericardium was used for intra-atrial rerouting. Surgical ASD was created in patients who had intact atrial septum. The anomalous PV was baffled into LA through the ASD. New PV channel was accomplished from the anomalous right upper PVs via the proximal SVC and the created tunnel in RA to LA. CFD analysis of both venous channels was performed with 4D Flow MRI.
Results: The median follow-up period was 10.7years. There was no early mortality and late death. Nobody required reoperation. All patients maintained sinus rhythm and supraventricular arrythmias didn't occur. The median blood flow velocity of the neo-SVC channel was measured 0.47 m/s, and that of the neo-PV channel was measured 0.41 m/s. Among the 6 patients who underwent CFD analysis, the straight and non-restrictive flow stream line was visualized in both pathways, and low wall share stress was detected in all regions.
Conclusion: Newly developed double-decker technique is useful alternative surgical procedure for PAPVR to the SVC. Late complications can be completely avoided by this technique. Growth potential of both channels are maintained. CFD analysis is an useful method for such a complicated hemodynamics evaluation.

P2980 - THE EFFECTIVENESS OF LASSO TECHNIQUE IN BILATERAL PULMONARY ARTERY BANDING

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Purpose: We evaluated the effectiveness of Lasso technique in bilateral pulmonary artery banding (PAB).
Methods: Lasso technique:A expected size of bougie is catched with Lasso of Gore-Tex suture CV-4. The CV-4 are passed through the tourniquet (4Fr. ATOM tube), preoperatively marked with 7-0 prolene at the edge of the tourniquet. The right and left pulmonary arteries are encircled by this CV-4. After tourniquetting, Hemoclips are applied to the CV-4 at the marked site. Hemoclips are added to adjust the PA constriction. Intraoperative epicardial echography is performed to adjust the velocity at the PAB site, aiming to 3.0-3.5 m/s. We performed retrospective review of 29 patients who underwent bilatelara PAB from 2000 to 2016 at our institution.
Result: Bilateral PAB was performed at the age of 7.2 ± 4.7 days. The velocity and PA circumference at the banding site was 3.2 ± 0.5 m/s and 8.9 ± 1.0mm in right, 3.3 ± 0.6 m/s and 8.9 ± 1.0mm in left respectively. Preoperative SpO2 were declined from 92.4 ± 4.5% to 83.2 ± 5.3% after bilateral PAB. No reoperation was required for migration of the PA banding. Reoperations for additional adjustment of the PA band were required in 4 cases due to excessive pulmonary blood flow or systemic desaturation. Each adjustment was performed easily by adding or removing the hemoclip. At the second stage operation, PAB was removed easily by cutting the CV4. In 5 cases, PA reconstructions were required to release PA stenosis at the banding site. PA index at the banding site was 198.6 ± 66.2(110.3 ± 41 in right, 88.3 ± 32.3 in left).
Conclusion: Lasso technique is attractive method in terms of feasibility, re-adjustability and PA growth.

P3010 - CAN MONOCUSP VALVES BE DISPENSED WITH IN YOUNGER PATIENTS WHILE DOING RVOT RECONSTRUCTIONS

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Background: Transannular patching (TAP) of the right ventricular outflow tract results in pulmonary insufficiency (PI) The objective of this study was to (i) compare early outcomes after TAP with monocusp valve reconstruction or TAP alone and (b) assess the

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mid-term results after polytetrafluoroethylene (PTFE) membrane monocusp reconstruction.
Methods: From 2014 through 2016, 62 patients (mean age, 2.3 + /- 5.0 years) underwent right ventricular outflow tract reconstruction with a TAP. Data was retrospectively obtained to compare clinical outcomes among two groups: patients receiving monocusp with TAP (group A, n = 21) and patients undergoing TAP without monocusp (group B, n = 41).
Results: Overall, there were 1 early and 4 late deaths (5/62; 8.1%). There was a significant reduction of the preoperative right ventricular outflow tract gradient postoperatively following TAP (96.2 vs 16.1; P < .0001). Postoperatively, the RV/LV pressure ratio was <0.5 in all groups. Mechanical ventilation time, fluid drainage duration and total ICU stay showed no significant difference between Groups A and B. The incidences of moderate or severe PI on discharge were 9.5% in Group A, 7.3% in Group B (P = 0.7). None of the patients underwent reoperation for monocusp replacement. There was one death secondary to infective endocarditis of monocusp valve (4.7%). Two (9.5%) patients had moderate right ventricular outflow tract stenosis across the monocusp patch (mean gradient, 36.5 + /- 20.3 mm Hg). These gradients showed a inverse linear correlation with the age of the patient.
Conclusions: Use of a PTFE-monocusp valve prevents immediate PI, though no significant improvement in early or mid-term clinical outcomes is noted. There is a significant risk of pulmonary embolization, infective endocarditis which should be considered before implanting a monocusp valve. There is an increased risk of residual PS especially in younger patients which may increase the risk of reoperations.

P3021 - THE ROLE OF PALLIATIVE SURGERY IN THE ERA OF THE CORRECTIVE SURGERY. ANALYSES OF RESULTS AND FUTURE PERSPECTIVE

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Objective: To evaluate the results in surgical procedures of univentricular hearts.
Patients and Methods: Retrospective analysis of the 42 patients operated from 2013 to 2016.
Results: Neonatal palliation (16 patients): Blalock-Taussig shunt: 7 cases; Norwood procedure: 5 patients; Banding 4 cases. BT shunt: age 23 days (2-40); Aristoteles 7.5. Medium stay at PICU 12 days (5-32) and hospital 32 days (10-97). 1 reintervention due tu acute thrombosis. No mortality. Norwood procedure: 5 patients. Age 29 (6-90) days. Aristoteles 20.8. Stay at PICU 35 (19-57) days and hospital 48 days. Morbidity: 1 case of inferior vena cava thrombosis. Mortality: 1 case due to refratory septic shock. All cases have reached the Glenn procedure. Glenn procedure: 11 patients (1 case associated to Damus-Kaye-Stansel procedure). Age: 8(4-135) months. Aristoteles 9. Stay at PICU 17 (3-90) days and hospital 27 (7-119 days). Morbidity: 1 case of phrenic nerve paralysis. Fontan procedure: 15 patients. Age 56 (40-113) months. Aristoteles 11. Stay at PICU and hospital: 7 (4-81) and 24 (14-90). Morbidity: 1 case of cardiac tamponade due to chylopericardium, 1 case of persistent pleural effusion (reintervention and embolization of thoracic duct and 1 case of thrombosis of inferior vena cava. No mortality. Overall mortality: 2.3% In one particular case, the use of a MRI 3D printed model redirect a case from a Glenn procedure to a biventricular heart.
Conclusions: Surgery of univentricular hearts means complex high risk procedures. Currently, they can be performed successfully and patients are able to reach the definitive palliation with the Fontan procedure. The use of new methods for accurate diagnostic could help patients to be reconsidered from single ventricule-surgery to biventricular surgery.

P3033 - ACQUIRED LEFT VENTRICLE TO RIGHT ATRIUM COMMUNICATION AFTER CARDIAC SURGERY

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Background: Left ventricular (LV) to right atrial (RA) communication is very rare congenital defect, but sometimes caused by complications of cardiac surgery, infectious endocarditis, trauma or myocardial infarction. Acquired LV to RA communication after surgery is rare complication but can be hazardous for the patient. Recently, awareness of this complication has been increased.
Methods: We analyzed medical records of 6 patients complicated by acquired LV to RA communication after cardiac surgery for congenital heart disease (CHD) at Chungnam National University Hospital (4) and others (2), retrospectively. We investigated the underlying CHD and surgical procedures. In addition, clinical manifestation, course and outcome of LV to RA communication were analyzed.
Results: Underlying CHDs were ventricular septal defect (VSD) (4), VSD, atrial septal defect and persistent ductus arteriosus (1) and transposition of great arteriosus with VSD(1). All patients performed autologous patch closure for VSD. All LV to RA communication were occurred postoperatively except one by infective endocarditis at patch closure region. The diagnosis was confirmed with transthoracic echocardiography. In addition, cardiac catheterization was done for shunt evaluation in two patients. Three patients underwent patch closure for LV to RA communication. In one patient, phrenic nerve palsy was complication by operation for LV to RA communication.
Conclusion: Acquired LV to RA communication is very rare and not well-known but significant complication after cardiac surgery for CHD. In many cases, the symptom and sign are not clear and variable. So, detailed and careful cardiac evaluation is needed to detect the LV to RA communication after cardiac surgery.

P3044 - ANOMALOUS ORIGIN OF THE CORONARY ARTERY FROM THE PULMONARY ARTERY DIAGNOSIS AND SURGICAL RESULTS IN 16 PAEDIATRIC PATIENTS OVER A 13 YEARS PERIOD

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Background: Anomalous origin of the coronary artery is rare, the origin of the left main coronary from the pulmonary artery (ALCAPA) is 0.25% of all congenital heart disease and even more rare is the origin of the right coronary artery from the pulmonary artery (ARCAPA). The aim of this study was to describe a single-

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centre experience with surgical repair of this condition and determine the midterm outcome.
Materials and Methods: We studied 14 patients (11 females) with ALCAPA and 2 ARCAPA (males) who underwent surgical correction between 2003-2016 in our institution. All patients underwent chest radiography, ECG, and echocardiogram. In 5 patients CT angiogram or angiography was also performed. All patients underwent surgical reestablishment of a dual coronary system with direct aortic reimplantation of the anomalous origin coronary artery into the aorta.
Results: The patients' mean age at presentation was 2.25 ± 4 years (range, 1mo–15 yr). Eleven were infants (age, <1 yr); of these 1 was neonate and 7 were <4 months old. Clinical presentation in ALCAPA patients was heart failure in 5 infants, failure to thrive in 2 infants, cardiogenic shock in 1 neonate, ventricular fibrillation in one child and incidental murmur in the remaining ones. All the infants aged <6mo had dilated cardiomyopathy. The 2 ARCAPA patients were diagnosed following incidental finding of a murmur. At the last follow-up (mean 6.7 + 3.6years, range 3-12 yr), all the patients were alive, left ventricular systolic function was normal (EF > 55%) and all were asymptomatic. Mitral valve (MV) repair, performed in 6 (38%) ALCAPA patients with ≥ moderate MV incompetence, resulted in mild incompetence at the last echo evaluation.
Conclusions: Although initial presentation can be severe, the outcome of this cohort's patients after successful direct reimplantation was favourable. Additional MV repair is commonly required. High level of suspicion is crucial for improving diagnosis.

TRANSPLANT/HEART FAILURE/PULMONARY HYPERTENSION

P1004 - OUTCOMES IN PAEDIATRIC POST MYOCARDITIC AND PRIMARY DCM

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We compared basal data and outcomes of children with primary DCM and myocarditis. All patients (0-18 yrs.), referred in 2000-2016 to a single University Hospital for LVEF < 55%, and/or LVEDD >2 Zscores forBSA, were reclassified as: 32/72 myocarditis (11/72 definite, 8/72 probable, 13/72 possible) or unlikely 40/72(DCM). Admittance to ICU and inotropes use were similar (16/32 and 14/32 M vs 13/40 and 10/40 DCM) (p = 0,1 and 0,3 respectively), as was the need for ECMO or VADs (6/32 M vs 4/40 DCM p = 0,3). LVEF at onset was significantly lower (p = 0,01) for M patients (29,6% vs 39,1%) while mean LVEDDz was comparable (4,1 ± 2,1 M vs 3,5 ± 2,6 DCM p = 0,3). 4 patients were lost to follow-up. 6/30 patients in the M group and 8/38 in the DCM group reached the primary end point of death or transplant(p = 0,21). LVEF was significantly lower in patients progressing to this primary endpoint (32 ± 11,7 vs 17 ± 8 p = 0,09 for M, and 44,8 ±17 vs 25 ± 9 p = 0,07 for DCM), independently on the LVEDDZ score (3,6 ± 2 vs 5,8 ±1 p = 0,6 for M, and 3,5 ± 2,3 vs 5,4 ± 2,1 p = 0,2 for DCM). Onset below 2 years was associated with death or transplant in the DCM group only (7/22 vs 1/18 p = 0,02 vs 3/18 vs 3/14 p = 0,5). Recovery to normal LV was observed in 17/68 patients, with a non significant trend (p = 0,18) for better outcomes in the M group (10/30 vs 7/38). Higher LVEF at diagnosis was associated with a higher chance of recovery in the M group (29,6 ± 14 vs 41,7 ± 6,9 p = 0,008 inM, 26 ±11,8 vs 38,9 ± 9,7 p = 0,11 in DCM). Hence, it is not possible to confirm a strikingly better outcome for paediatric M patients.

P1048 - SCHOOL INTERVENTION THE MISSING LINK IN FOLLOW UP CARE

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Background: Children with congenital heart disease (CHD) are at risk for neurodevelopmental delays and differences due to changes in blood flow affecting brain development. Children with CHD score lower in hand-eye coordination, fine and gross motor skills, social-emotional functioning, and language development than their typically developing peers. Frequent attention problems and behavioral concerns are additionally reported. The Children's Hospital of Wisconsin, Herma Heart Center established their School Intervention Program (SIP) in February 2015 to address these educational risk factors. The SIP conducted a pilot study designed to promote academic success of pediatric heart transplant patients by providing individualized assessments and implementing intervention strategies in collaboration with their school. Services expanded to support all areas of pediatric cardiology in May 2016. No data currently exists on the benefits of structured school intervention as a comprehensive support service for pediatric cardiology patients.
Materials and Methods: We performed a retrospective, single center analysis of pediatric cardiology referrals to the SIP from 2/2015 to 10/2016. Referral rates, assessment results and interventions were reviewed and catalogued for descriptive analysis.
Results: Of 63 total school-age patients referred for enrollment, 57 (90.5%) required interventional follow-up. Special education support was the most common school need, while bullying was the most common secondary concern. General fear and lack of knowledge leading to poor communication, low expectations for the student resulting in low achievement and motivation, and an overall identification of missed opportunities for utilizing care coordination were significant trends. Through structured intervention facilitation, 54% of school concerns were resolved with no further action needed, while 40% remain on-going with improvements reported.
Conclusions: Integration of a SIP as part of multidisciplinary follow-up care provides standardized structure to address these known educational and developmental needs. Future research on programmatic correlation to medical outcomes and overall quality of life is needed.

P1111 - THE AORTIC PERFUSION SCORE A NOVEL SCORING SYSTEM TO PREDICT DEATH OR TRANSPLANT IN CHILDREN WITH PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM

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Background: Pulmonary atresia with intact ventricular septum (PAIVS) is a unique congenital malformation. Various factors can impact the selection of appropriate palliative strategy. We developed the aortic perfusion score (APS), a novel scoring system based on anterograde coronary perfusion with the aim of being able to identify patients at risk for death or transplant.
Material and Methods: A retrospective study of patients at our institution was conducted. Patients were included if an initial catheterization was done prior to any intervention. Each patient was assigned an APS. In this system, each of the named coronaries (left anterior descending, left circumflex, right main, and posterior descending) had a maximum of 100 points they could be assigned.

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If the entire length of the coronary was being supplied via the aortic root then the coronary would receive 100 points, if only a percentage of it were receiving supply from the aortic root then it would receive that many points. If there was to-fro flow in the coronary due to VCCs then the coronary artery's score was halved. These were then summated for a maximum score of 400. ROC analysis was done to determine a cutoff point predictive of a composite endpoint of death or transplant.
Results: A total of 64 patients were included in the analysis with 10 reaching the endpoint of death or transplant. An APS of 227.5 predicted the endpoint with a sensitivity of 90% and a specificity of 83%.
Conclusion: The APS is a scoring method which can be used to predict a composite endpoint of death or transplant in patients with PAIVS and may be helpful in selecting patients that should be listed for transplant. Additional multicenter studies may help further refine the scoring system.

P1165 - PROGNOSTIC VALUES OF HEART RATE AND OXYGEN SATURATION IN PATIENTS WITH PULMONARY ARTERIAL HYPERTENSION BEFORE AND AFTER SIX MINUTE WALK TEST

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Objective: This study aims to determine the prognostic values of heart rate (HR) and transcutaneous oxygen saturation (Sat) which were measured before six minute walk test (6MWT), at the end of 6MWT and 5 minutes after 6MWT in children who are undergoing pulmonary arterial hypertension specific treatment (PAH-Rx).
Methods: 29 children (age 7 -17 years) were evaluated. Sat and HR were measured before 6MWT, at the end of 6MWT and 5 minutes after the completion of 6MWT (Sat0, Sat1, Sat2; HR0, HR1, HR2 respectively). HR increase was defined as HR1 minus HR0 and decrease in Sat was defined as Sat0 minus Sat1.
Results: The mean age was 129 ± 45 months and the mean follow-up time was 58 ± 40 months. The mean pro-BNP concentrations did not change significantly before and after the PAH-Rx (946.4 ± 1754.8 pg/ml vs 917.6 ± 1920 pg/ml $p = 0.11$). However, the mean 6MWT distance increased significantly after the PAH-Rx (400.2 ± 107.8 m vs 436.2 ± 119.2 m, $p = 0.012$). The 6MWT distance was significantly longer and HR1 was significantly higher in survivors (453.3 ± 96.5 m vs 250 ± 135.2 m, $p = 0.025$ and 122.8 ± 18.4 /min vs 94.3 ± 19.1 /min, $p = 0.034$). HR1 increased significantly but Sat1and Sat2 decreased significantly after PAH-Rx ($p = 0.017$, $p = 0.03$ and $p = 0.017$ respectively). While there was no significant correlation between WHO-FC and Sat1 before treatment, WHO-FC and Sat1 correlated negatively after treatment ($r = -0.435$, $p = 0.021$). Although WHO-FC and pro-BNP did not correlate before treatment, they correlated positively after treatment ($p = 0.0001$). There was no association between survival and HR increase whereas survival was positively associated with the decrease in Sat. If the decrease in Sat was 14.6%, sensitivity increased to 100% and specifity was 58.3%.
Conclusions: PAH is a chronic disease which continues to progress despite the administration of specific therapy. The measurement of Sat with pulse oximeter during 6MWT might have prognostic importance in the follow up of children undergoing PAH-Rx.

P1209 - CORRELATION OF HEART FAILURE SEVERITY AND N-TERMINAL PRO-BRAIN NATRIURETIC PEPTIDE LEVEL IN CHILDREN AGED ONE MONTH TO 14 YEARS

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Background: Heart failure affects the morbidity and mortality in children with heart disease. There is no single specific test to diagnose heart failure. The modified Ross Reithmann scoring system has been used to classify heart failure severity, but it is limited due to its subjectivity. The N-terminal-pro brain natriuretic peptide (NT-proBNP) is secreted by the ventricles during heart failure.It has been suggested as a possible marker for diagnosing heart failure.
Objective: To investigate the correlation between heart failure severity and plasma NT-proBNP concentration in children aged one month to 14 years.
Methods: A cross-sectional study was performed in Pediatric Department of Mohammad Hoesin Hospital Palembang, Indonesia from August to October 2016 on children with congestive heart failure, aged one month to 14 years. Heart failure severity was assesed using the modified Ross Reithmann scoring system. Plasma NT-proBNP measurements were done in all subjects. Statistical analysis was done by Spearman's test.
Results: Subjects median plasma NT-proBNP concentration was 1.703 pg/ml (range 310 - 9.000 pg/ml). The NT-proBNP level and severity of heart failure had a significant positive correlation ($r = 0,87; P < 0,001$). The NT-proBNP minimum levels in subjects with mild, moderate and severe heart failure were 310 pg/ml, 1.251 pg/ml, and 2.610 pg/mL respectively.
Conclusion: Plasma NT-proBNP level has a significant, positive correlation with the severity of heart failure in children. As such NT-proBNP level may be useful as a biochemical marker for the diagnosis and grading of severity of heart failure in children.

P1212 - COMPARISON OF CLINICAL PROFILES BETWEEN PATIENTS WITH PROTEIN LOSING ENTEROPATHY WITH AND WITHOUT FONTAN CIRCULATION

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Background: Protein losing enteropathy (PLE) is one of life-threatening complications in patients after Fontan operation. However, PLE sometimes occurs in patients with congenital heart disease (CHD) after biventricular repair (BVR). Objectives: Our study was conducted to compare clinical profiles of PLE patients with and without Fontan circulation.
Materials and Methods: We retrospectively reviewed clinical charts of all our postoperative CHD patients with PLE.
Results: We had a total of 42 PLE patients (BVR in 14, Fontan in 28). Postoperative follow-up period was significantly shorter in the Fontan group than in the BVR group (14 ± 2 years vs. 8 ± 1 years, $p = 0.02$) despite there were no difference in age at the onset of PLE between the two groups. Prevalence of clinically relevant arrhythmias was equivalent between the two groups. Regarding hemodynamics, although there were no differences in cardiac output or central venous pressure at the onset of PLE between two groups, percentage of treatable structural lesions (valve regurgitation and stenotic lesions) by non-pharmacological interventions

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was higher in the BVR group than in the Fontan group (93% vs 50%, $p < 0.01$). During the follow-ups, 2 BVR and 8 Fontan patients died. There were no differences in the 5 to 10-year survival after the onset of PLE (81% vs 81%, 81% vs 66% for the BVR and Fontan, respectively). However, the complete remission rate from PLE was higher in the BVR group than in the Fontan group (38% vs. 7%, $p = 0.02$).
Conclusions: Mortality of CHD patients with PLE was substantially high regardless of the postoperative hemodynamics, BVR or Fontan physiology. BVR patients might have a chance of remission from PLE because of the possible treatable responsible lesions.

P1246 - FROM PATIENT TO PROFESSIONAL A TALE OF TWO PERSONAL JOURNEYS IN A PAEDIATRIC HEART TRANSPLANT CENTRE

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Background: Heart transplantation is an accepted therapy for children with end-stage heart disease. The Royal Children's Hospital, Melbourne is the nationally designated paediatric heart transplant service for all children aged 0-18 years.
Methods: This abstract presents the experiences and challenges for two former patients who have returned to RCH, CK as a PICU nurse and RP as an adolescent transition support worker. CK was diagnosed with dilated cardiomyopathy at age 14 and relocated to Melbourne from South Australia, for VAD placement. She underwent transplantation several months later. CK graduated from nursing and works in the RCH PICU. RP was diagnosed with LVNC at 8 weeks of age. She relocated from the Northern Territory at age 7 and underwent heart transplantation. RP now works at RCH assisting young people with chronic illness, transitioning to adult care.
Results: 70% of all transplants at the RCH are performed from circulatory support and the number of transplants has increased to 12/year. Paediatric donor utilisation is 71%, with excellent post-transplant outcomes. Both RP and CK can relate to all aspects of the transplantation journey. For CK this involves a deeper understanding of the hopes and fears of families requiring ICU or circulatory support. For RP this involves understanding the challenges facing adolescents with a chronic illness, preparing to transition into adult life. This knowledge allows RP and CK to care for these patients at a higher level. Protective strategies include not discussing their personal history, diversifying their academic and clinical roles and setting personal goals to promoting physical and mental health.
Conclusions: Improving transplant rates and outcomes has led to more transplant patients working in the healthcare sector. This unique background enables them to provide a personalised level of professional support to transplant recipients, and to offer guidance in managing chronic illness with resilience.

P1267 - TIPS AND PITFALLS IN TRANSPLANT AFTER FONTAN

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Background: Heart transplantation after Fontan completion poses a unique surgical challenge. Twelve patients are presented, stressing the technical hints performed in the five anastomoses to match the graft in the recipient.
Methods: Data are collected from ten Fontan and two takedown patients along 4 consecutive years. Age (9 years), weight (30 Kg.) and time interval between Fontan and transplant (3 years) are presented as median. Extra cardiac conduit (size 18/20) was implanted in 8 patients, whereas atriopulmonary connection was found in three and lateral tunnel in one. Three patients developed protein losing enteropathy. Ten stents have been previously deployed in left pulmonary artery (6), inferior vena cava (3) and right pulmonary artery (1). One patient was on Levitronix for two weeks before transplant.
Results: The five anastomoses underwent some changes. Left atrium once (enlargement with recipient both atria), aorta three times (hemi-arch repair), superior vena cava three times (one case with double superior vena cavae), pulmonary branches eight times (hilum to hilum pasty with donor´s aorta/pericardium patch after thorough stent removal) and inferior vena cava eight times (conduit sleeve anastomoses). Follow-up was complete for a median of 35 months (range 1-48) One patient died. ECMO was needed in four cases for pulmonary hypertension. Three patients had collateral vessels occluded in the cath-lab and stents placed in superior vena cava (1) and aorta (1). Protein losing enteropathy resolved in two children. Interestingly, our latest patient was on systemic assist device before transplant and right assistance (ECMO) afterwards.
Conclusions: Transplant in Fontan patients is actually challenging. Hints in every of the five proposed anastomoses must be anticipated, including stents removal. Extra tissue from the donor (innominate vein, aortic arch, pericardium) is strongly advisable. ECMO for right ventricular disfunction was needed in one third of cases. Overall results can match other transplants cohorts.

P1270 - INITIAL IMPLANTATION OF LEVITRONIX VENTRICULAR ASSIST DEVICE PRIOR TO CONVERSION TO BERLIN HEART; A COMPARITIVE STUDY

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Background/Hypothesis: Scarcity of donor hearts has increased the demand for reliable mechanical support. We are one of two centres in the United Kingdom who use Ventricular Assist Devices (VAD) as a bridge to transplant. In previous years the most common VAD used was a BERLIN HEART; however, due to visible fibrin and clot deposition and risk of systemic embolism we required numerous ventricle changes. The cost of this, led us to change our policy in that we now use the LEVITRONIX centrifugal VAD from implantation, therapeutic heparinisation and platelet inhibition is achieved prior to conversion to a BERLIN HEART device. Several implications have been recognised for a child being on LEVITRONIX opposed to a BERLIN HEART.
Materials and Method: We hypothesised that patients <10 kg would have the highest incidence of clot deposition requiring ventricle change and that using LEVITRONIX in this group would lead to fewer pump changes at the expense of reduced morbidity. Following a comparative study, using a representative sample of our VAD patients since 2010, patients selected met the following criteria: <10 kg, supported for > two weeks on mechanical support, with a minimum of seven days on a BERLIN HEART with the ventricle size 10-15mls. This information is derived from Freeman Hospitals ECMO/VAD as a bridge to transplant database.
Results: Comparison between patients implanted with the BERLIN HEART device against patients implanted with

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LEVITRONIX prior to conversion to BERLIN HEART and looking at the number of ventricle changes carried out.
Discussion/Conclusion: Our study is not representative of all VAD patients however it demonstrates that stabilising children on LEVITRONIX leads to more manageable coagulation when converted to a BERLIN HEART. There's an increased risk to patients on mechanical support with unstable coagulation therefore it is important that it is stabilized as soon as possible to reduce risk of morbidity.

P1271 - LEFT VENTRICULAR END DIASTOLIC DIAMETER OBTAINED DURING FOLLOW UP ECHOCARDIOGRAPHY PREDICTS MORTALITY IN CHILDREN WITH HYPERTROPHIC CARDIOMYOPATHY

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Background: In children with hypertrophic cardiomyopathy (HCM), the septal and free wall thickness at diagnosis predicts mortality. However, few studies have reported the clinical relevance of left ventricular measurements during follow-up echocardiography. We aimed to identify echocardiographic parameters that predict mortality during the follow-up of children with HCM.
Materials and Methods: After searching the echocardiography database for examinations performed from March 2013 to September 2016, we included 21 patients (11 boys) with HCM who were younger than 20 years of age when examined. Echocardiographic data from the latest examination were included for the analysis. Linear measurements of the left ventricle were standardized using z-scores based on body surface area. The echocardiographic data were compared for survivors and non-survivors.
Results: Ten patients had Noonan syndrome, 2 had LEOPARD syndrome, and and 2 had familial history of HCM. The median age at echocardiographic evaluation was 7.1 (0.4-19.1) years. During a median follow-up period of 4 (1-41) months, 5 deaths (2 due to heart failure and 3 sudden deaths outside the hospital) occurred at a median of 4 (1-9) months after echocardiographic examination. Left ventricular end-diastolic diameter z-score was significantly larger in non-survivors than in survivors (1.2 ± 0.6 vs. -1.5 ± 1.9, $p < 0.05$). Non-survivors showed significantly smaller left ventricular fraction shortening z-score than survivors (-0.6 ± 5.0 vs 4.8 ± 4.3, $p < 0.05$). There were no significant differences in septal and free wall thickness z-scores between the groups.
Conclusions: A less compressed left ventricular cavity size is associated with mortality, which suggests that congestive heart failure is a pivotal contributor to death in children with HCM. In the follow-up of children with HCM, a left ventricle with a preserved internal diameter and decreased contraction despite concentric hypertrophy might imply poor outcome, and warrants further evaluation and therapeutic intervention.

P1361 - GENETIC ETIOLOGIES OF ACUTE HEART FAILURE IN PRE ADOLESCENTS

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Background/Hypothesis: There are a variety of causes of acute heart failure in pre-adolescent patients including myocarditis, genetic/metabolic conditions, and congenital heart defects. In cases with a structurally normal heart and a negative personal and family history, myocarditis is often presumed to be the cause, but we hypothesize that genetic causes are a significant portion of these cases.
Materials and Methods: We reviewed our cases of patients 12 years-old and younger who presented with acute heart failure and underwent genetic testing from 2008 to 2016.
Results: We present five cases of children from four families presenting with acute heart failure, who were determined to have a genetic etiology. Myocarditis was originally presumed to be the cause in all four families. First, a 12 year old female presented with acute heart failure. A cardiomyopathy panel revealed she has a pathogenic variant in TNNT2 (p.Lys210del). Secondly, a 3 month-old presented with dyspnea and cardiomegaly. Genetic testing showed two pathogenic variants in IDUA (p.Trp402* and p.Gln70*) in trans which have been shown to cause Hurler syndrome. In the third family, a pair of siblings presented with heart failure and underwent heart transplant during infancy. Limited genetic testing on the proband was uninformative, but when the second child presented, exome sequencing revealed two pathogenic variants in ALMS1 (c.1111_11134del19 and c.1794_1801dup8) in trans which cause Alstrom syndrome. Finally, a 2 month old presented with cardiomegaly. A cardiomyopathy panel revealed a "likely pathogenic" frameshift in TTN (p.Pro28826fs). Significantly, there was no history of dysmorphic features or prior cardiac disease in any of the cases and only a family history in the third family.
Conclusions: Genetic etiologies should be considered in children with acute heart failure even if there is no significant history to suggest a genetic cause.

P1380 - ABSTRACT FOR TRAFFIC LIGHTS SYSTEM TOOL FOR PAEDIATRIC INHERITED CARDIAC CONDITIONS PATIENTS PRODUCED BY NURSE SPECIALISTS FOR PATIENTS AND THEIR FAMILIES

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Background/Hypothesis: NHS England (2016) recognises that Early Warning Scores (EWS) aid identification of children at risk of clinical deterioration. NICE (2007) have strongly advised that EWS should be utilised for all inpatients. However, there is limited evidence to support the use of EWS within outpatient care for patients with chronic conditions. Literature recommends teaching patients symptom monitoring skills to recognise elevated heart failure risk. It is imperative that patients are able to identify signs of deterioration and seek appropriate help. Use of a symptom management tool can aid timely identification and management of symptoms, and access to relevant health services (Vincent and Mutsch, 2015).
Materials and Methods: A Traffic Light System is being trialled within the Paediatric Inherited Cardiac Conditions (ICC) Service at Royal Brompton Hospital, developed with the view to act as a EWS for patients to use once discharged post inpatient care. It aims to; identify the severity of patient symptoms, relieve patient anxieties and support patients in taking appropriate actions in response to symptoms.

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Results: Feedback from the patients and families has yet to be collected in relation to the utilisation of the Traffic Light System. A questionnaire will be distributed to families who have used the tool, for the purposes of collecting and analysing data. This will assist in examining the effectiveness of the tool, and will ascertain whether or not it is beneficial for this patient group. Further improvements and developments can then be made in light of the findings.
Conclusions: Working with patients with chronic conditions involves a considerable amount of time supporting them with symptom advice and management. The Traffic Light System aims to empower patients to recognise symptoms, track them, identify early warning scores and for the patient/parents to act appropriately, is potentially beneficial for the patients and also for service development.

P1381 - MUSCLE STRENGTH IN CHILDREN WITH PULMONARY ARTERIAL HYPERTENSION IS DECREASED

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Background: Pulmonary arterial hypertension (PAH) is a rare, severe disease of the pulmonary arteries and has a poor prognosis. PAH is characterized by exercise intolerance due to impaired cardiovascular function. Recently, muscle strength was shown to be decreased in adults with PAH and may thus play a role in the exercise intolerance. To date, data in children are lacking. We aimed to assess whether muscle strength is decreased in children with PAH and to explore its correlation with physical functional parameters.
Methods: In this prospective study, eligible children with PAH aged 4-18 years who visited the Dutch National Referral Center for Pulmonary Hypertension in Childhood between October 2015 and March 2016 were included. Peripheral skeletal muscle strength was evaluated using maximum voluntary isometric contractility (MVIC) of the elbow flexor and extensor, hip flexor, knee extensor and grip strength. The contribution of muscle strength to motor proficiency was evaluated with the Bruininks-Oseretsky test of motor proficiency – 2nd edition (BOT-2). These measurements were compared with age and sex matched reference values and correlations with 6-minute walk distance (6MWD) and World Health Organization Functional Class (WHO-FC) were assessed.
Results: In total, 18 children were included. Peripheral skeletal muscle strength and BOT-2 were significantly decreased compared to mean reference values. MVIC of the elbow extensor, grip strength and BOT-2 significantly correlated with 6MWD ($r = 0.688$, $p = 0.002$; $r = 0.587$, $p = 0.017$ and $r = 0.520$, $p = 0.032$, respectively). Only BOT-2 correlated with WHO-FC ($r = -0.602$, $p = 0.010$).
Conclusion: Peripheral skeletal muscle strength and BOT-2 scores of children with PAH were significantly decreased compared to healthy references and correlated with 6MWD and WHO-FC. This decreased muscle strength may contribute to decreased functional capacity in children with PAH. The pathophysiologic mechanisms of this reduced peripheral muscle strength in children with PAH remain to be clarified.

Table 219. P1381 Table 1. Patient and disease characteristics at moment of muscle strength tests of the total study population (N = 18)

	All patients (n = 18)
Female (%)	12 (67%)
Age at tests (yrs)	14.0 (9.9 – 16)
Age at diagnosis (yrs)	6.2 (2 -12.2)
BMI (kg/m2)	18.2 ± 3.4
Diagnosis	
IPAH/HPAH	14 (78%)
PAH-CHD	3 (17%)
PAH-CTD	1 (6%)
WHO-FC	
I	4 (22%)
II	10 (56%)
III	4 (22%)
IV	0
6 MWD (m)	430 ± 82.3
NT-proBNP (ng/l)	131 (48.8 – 247.3)

BMI, body mass index; IPAH, idiopathic pulmonary arterial hypertension; HPAH, heritable pulmonary arterial hypertension; PAH-CHD, pulmonary arterial hypertension associated with chronic heart disease; PAH-CTD, pulmonary arterial hypertension associated with congenital tissue disease; WHO-FC, World Health Organization Functional Class; 6 MWD, six minute walking distance. Data are presented as mean (± standard deviation), median (interquartile range) or number (percentage).

P1391 - LONG TERM BENEFITS OF INTRAVENOUS EPOPROSTENOL IN IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION; A PERSONAL EXPERIENCE

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Introduction: In the past 15 years, medical therapy for patients with idiopathic pulmonary arterial hypertension (IPAH) has improved outcomes, including functional status and quality of life. Intravenous Epoprostenol is initiated when there has been an inadequate response or clinical deterioration with oral therapy, prior to consideration of lung transplantation. Despite these advances, patients with IPAH have considerable morbidity with many life altering challenges.
Method/Results: TD was diagnosed with IPAH in 1995 at the age of 6, presenting with symptoms of syncope, dizziness and shortness of breath. At that time there were no effective therapies available in Australia and the median life expectancy for affected children was less than one year from diagnosis. After a medical work-up she was commenced on Diltiazem with an initially good result, but several years later developed symptoms of heart failure. She improved somewhat with the addition of Bosentan in early 2001, but remained highly symptomatic. In late 2001 she was placed onto intravenous Epoprostenol via centrally inserted Hickman catheter and placed onto the wait list for lung transplantation. Within a few months the changes were profound; there were no further syncopal episodes, her exercise tolerance increased and she was able to return to school. Although not symptom free, TD now faces the challenges of her disease and the side-effects of her medication in such a way that she exceeds the expectations of a patient with IPAH. With strict medication management, self-observation and attention to detail, TD is able to work as a Clinical

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Nurse Specialist in Oncology, has a passion for travel and has explored over 20 countries, including 3 months visiting Northern Europe.
Conclusion: The use of intravenous Epoprostenol combined with appropriate self-care has allowed TD to lead a busy and independent life over many years.

P1396 - MODELING CARDIAC REGENERATION INDUCED BY RIGHT VENTRICULAR PRESSURE LOAD IN INFANT RATS

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Background: Right ventricular (RV) pressure load plays a major role in the development of RV failure in congenital heart diseases (CHD). Recent studies reveal a regenerative capacity of cardiomyocytes that can be addressed to find new treatment targets. We hypothesize that regeneration and proliferation plays a cardioprotective role in CHD that depletes progressively during childhood. Comprehension of these mechanisms could provide new insights to aid the prevention or treatment of RV failure in CHD. We developed a rat model of progressive RV failure during infancy due to pressure load (PL) to study the role of proliferation and regeneration in the heart.
Materials and Methods: Wistar rat pups (30-45 g) were subjected to PL (n = 10) or sham surgery (n = 7) and studied during childhood (~8 weeks). We reported symptoms of RV failure (e.g. bodyweight, dyspnea, pleural effusion/ascites) and performed analysis of functional and structural adaptation by means of sequential echocardiography and immunohistochemistry.

Table.

	2 weeks		4 weeks		8 weeks	
Weights	**SHAM (n = 7)**	**PL (n = 10)**	**SHAM (n = 7)**	**PL (n = 10)**	**SHAM (n = 7)**	**PL (n = 10)**
Bodyweight (grams)	110 ± 5	81 ± 4*	195 ± 8	126 ± 5*	293 ± 4	199 ± 12*
RV/(LV + IVS) ratio	#	#	#	#	0.78 ± 0.08	1.87 ± 0.24*
Clinical parameters						
Dyspnea	0%	0%	0%	0%	0%	80%*
Pleural effusion/ascites	0%	0%	0%	0%	0%	50%*
Echocardiography						
PL gradient (mmHg)	5 ± 0	51 ± 3*	6 ± 1	67 ± 6*	6 ± 0.1	87 ± 3*
TAPSE (mm)	2.5 ± 0.1	1.9 ± 0.2	3.2 ± 03	2.0 ± 0.3*	3.4 ± 0.3	2.7 ± 0.4*
CO (ml/min)	78 ± 10	31 ± 3*	133 ± 23	55 ± 10*	144 ± 16	68 ± 10*
EI systolic	1.0 ± 0.1	1.3 ± 0.1	0.9 ± 0.0	1.5 ± 0.1*	1.0 ± 0.2	1.8 ± 0.1*
EI diastolic	1.1 ± 0.0	1.3 ± 0.1	0.9 ± 0.0	1.5 ± 0.1*	1.0 ± 0.0	1.6 ± 0.1*
TI (present)	0%	60%*	0%	90%*	0%	100%*
Immunohistochemistry						
RV Ki67 (positive nuclei)	#	#	#	#	4.3 ± 0.6%	8.6 ± 0.6%*
RV CCSA (um^3)	#	#	#	#	320 ± 23	697 ± 26*
RV Masson's Trichrome	#	#	#	#	0.6 ± 0.2%	5.8 ± 1.1%*

Supplemental table. PL = pressure load. RV = right ventricle. LV = left ventricle. IVS = interventricular septum. TAPSE = tricuspid annular plane systolic excursion. CO = cardiac output. EI = eccentricity index. TI = tricuspid insufficiency. CCSA = cardiomyocyte cross-sectional area. Values are ± SEM, * = < 0.05 vs. SHAM. # = measurements were performed only after sacrifice.

Results: Severity of PL was confirmed by a clinical pattern characterized by failure to thrive, dyspnea and pleural effusion (see data in supplemental table). Chronic RV PL lead to decreased cardiac output and tricuspid annular plane systolic excursion after 2 weeks and septal bulging after 4 weeks. At sacrifice, after 8 weeks, RV hypertrophy and increased fibrosis were observed, both signs of pathological remodeling. However, in these ventricles, loaded during infancy, also increased cardiomyocyte proliferation, defined as cardiac cell-cycle activity (defined by Ki67) occurred.
Conclusion: This model of chronic RV PL in infant rats induces failure to thrive and RV failure, resembling CHD. The increased cardiac cell-cycle activity suggests a regenerative potential of the heart. This model can now be used to identify which cardiac cells can regenerate in response to pressure load and how this potential can be stimulated to prevent or treat RV failure in patients with CHD.

P1401 - CONTEMPORARY SURVIVAL OF PATIENTS WITH PULMONARY HYPERTENSION ASSOCIATED WITH CONGENITAL COMBINED AND COMPLEX SYSTEMIC TO PULMONARY SHUNT

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Background: Combined systemic to pulmonary shunt and complex shunt such as atrioventricular septal defect, truncus arteriosus, transposition of great arteries, single ventricle allow unrestricted flow to pulmonary circulation resulting in rapid progressive pulmonary arterial hypertension (PAH). Advanced management of PAH has been reported. However, cohort data in this anatomic-pathophysiologic group is still limited.
Objective: To compare the hemodynamic characteristics and survival of PAH associated with combined and complex shunt lesions.
Methods: Records of patients who had hemodynamic confirmation by cardiac catheterization of PAH with combined and complex shunt in 1995 – 2015 were reviewed. Patients who had single ventricle following any staged operation were excluded. Treatment and clinical at a recent follow-up were recorded. Mortality risk was performed using multivariate analysis. Kaplan-Meier curve were constructed and compared between the lesions.
Results: At a median follow up of 6.3 years (1-20.6 years), 181 patients (49% male) were reviewed. The median age of diagnosis was 4 years (0.4-69 years). There were 103 patients with combined shunt and 78 patients with complex shunt. Defect closures were performed in combined shunt more than complex shunt (77% vs 45%; $p < 0.01$) at the median age of 2.5 and 7.3 years, respectively. Comparing between groups, persistent PAH after correction occurred 38.7% and 28.5% and mortality rate was 7.8% and 14.1%. Survival rates of patients with combined shunt at 1, 5 and 10 years following diagnosis were 96%, 96% and 83.6% whilst complex shunt were 94.7%, 87.8% and 82.2%, respectively. Significant risk of mortality was baseline PVR >8 WU•m2 (p 0.03).
Conclusion: PAH associated with combined and complex shunt has a modest long term survival. Following defect correction, PAH persists in 1/3 of patients, despite deliberately selected patients. However, baseline PVR may be a useful marker in identifying patients requiring further aggressive medical therapy.

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P1420 - PULMONARY HYPERTENSION IN PEDIATRIC PATIENTS DUE TO LEFT HEART'S DESEASE

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Background/Hypothesis: Little is known about the evolution of pediatric patients (P) with pulmonary hypertension (PH) due to the left heart's disease (LHD) [PH-LHD]. We hypothesized that they are underdiagnosed and would benefit from timely treatment.
Objetives: To evaluate incidence, clinical and hemodynamic characteristics of the PH-LHD, response to treatment and its long term evolution.
Materials and Methods: After retrospective analysis of 132 clinical records of pediatric P with PH, 15 with PH-LHD were checked, evaluating sex, age and functional class (FC) at the moment of diagnosis, comorbidities, performed procedures, medical treatment used and survival. Left heart function was assessed by ejection fraction and tissue Doppler measures and hemodynamic assessment (pulmonary vascular resistance- PVR, diastolic pressure gradient-DPG) was performed to classified HP-LHD in isolated poscapillary and combined postcapillary and precapillary. Variables were analyzed by chi square test and Fisher's exact test.
Results: 11% of incidence PH-LHD. Average age 4 years (0.5-12), male (66%), average follow-up time 3 years (0.6-7). 55%, 35% and 10% were in FC II, III and IV respectively at the time of diagnosis. 12% of the P presented genetic syndromes. LHD was 8%/92% congenital obstruction at the inflow/outflowtract of the left heart respectively, 20% combination of both. 78% presented left ventricle sisto-diastolic dysfunction. 38% of PH-LHD was combined ($p < 0.05$). 13 P (86%) ($p < 0.05$) underwent heart surgery, 33% ($p < 0.05$) of them underwent Interventional catheterization during the follow-up time, 47% ($p < 0.05$) were then treated with vasodilators with 28% and 15% average decrease in the previous values of GDP and PVR respectively ($p < 0.05$). They went from FC III-IV to I-II, without late deaths. The untreated group remained on poor FC with two deaths ($p < 0.05$).
Conclusions: The incidence of PH-LHD is not negligible and timely vasodilator therapies improve FC and survival of the pediatric P.

P1443 - RESTRICTIVE CARDIOMYOPATHY IN CHILDHOOD RISK FACTORS AND OUTCOMES

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Restrictive Cardiomyopathy (RCM) accounts for 2.5 to 5% of all idiopathic cardiomyopathies in childhood. RCM is a serious, progressive disease, with a mortality rate of up to 50% within the first two years after diagnosis. Children with RCM develop progressive heart failure, arrhythmias, stroke, and sudden death.
Objective: to assess risk factors and progression in a group of patients diagnosed with RCM in a public institution.
Material and Method: In a retrospective analysis of a 36-patientcohort diagnosed with RCM, who were admitted to the Cardiomyopathy Unit at our hospital, clinical, echocardiographic, and hemodynamic findings, along with anatomical pathology, survival data,and cause of death were assessed.
Results: The overall survival rate was of 86 months (95% CI: 59 to 113). The multivariate analysis indicated that risk factors with the poorest prognosis were:the Shortening Fraction less than 28% (p 0.002), the LA:Ao ratio > to 2.5 (p 0.04), the S/D ratio less than 1 and the DT less or equal to 100 ms(p 0.05), and the age. The survival rate in children under 5 years of age was 43 months (95% CI: 35 to 57),whereas children above 5 years of age had a mean of 104 months (95% CI: 69 to 139). The other assessed variables, such as the types of cardiomyopathy, arrhythmias, and other echocardiographic parameters, were not significant. Causes of death were: progressive heart failure (16p), sudden death (2p).Nine patients were transplanted, whereas three patients were enrolled on the waiting list.Four out of the nine transplanted patients had a PVR of 6 WU/M with severe heart failure and were placed on VAD (Berlín Heart) as a bridge to transplantation.
Conclusions: Younger patients were the ones with the lowest survival rate. The Shortening Fraction, the LA:Ao ratio, the DT and S-D ratio were predictive risk factors.Sudden death caused by arrhythmias should be considered in asymptomatic patients.

P1446 - ROLE OF KALIISTATIN AND PRO ANTI INFLAMMATORY CYTOKINES IN PULMONARY HYPERTENSION

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Introduction: Kallistatin, serin proteinase inhibitor exhibits its properties in inhibiting inflammation, oxidative stress and apoptosis, angiogenesis. Severe pulmonary hypertension caused by left to right shunt congenital heart disease and Eisenmenger syndrome is progressive obliterative vasculopathy; the pathogenesis endothelial dysfunction and function of ion channels, calcium homeostasis, changes in platelet and endothelial function, intravascular thrombosis proliferation reactivity increased vascular inflammation and remodeling. In this study, we aimed to show the possible relationship between kalistatin levels, pro-inflammatory TNF-alfa and anti-inflammatory cytokine IL-10 levels in pulmonary arterial hypertension.
Methods: The study was performed in Erciyes University Medical Faculty Hospital. Patient group was formed from pulmonary hypertensive patients (Eisenmenger syndrome and primary Pulmonary Hypertension). We enrolled the patients with pulmonary arterial hypertension diagnosis that confirmed by the angiocardiography in our center. Pulmonary arterial hypertension is defined as mean pulmonary arterial pressure greater than 25 mmHg. Serum kallistatin, TNF-alfa, IL-10, NtProBNP levels were studied in each patient.
Results: The study included total 50 patients:25 patients in pulmonary hypertension group (16with Eisenmenger syndrome, 9 with primary pulmonary arterial hypertension). Control group consisted 25 patients with innocent murmur. Median value of kallistatin in pulmonary hypertension group was 1.42(1.0-1.58), control group 2.27(1.57-3.38). Serum levels of kallistatin were significantly lower ($p < 0.05$) in pulmonary hypertensive patients. Negative correlation was detected between mean pulmonary arterial pressure and serum kallistatin levels. However no correlation was found for proinflammatory cytokine TNF-alfa and anti-inflammatory cytokine IL10.
Conclusion: Anti-inflammatory and anti-apoptotic features of Kallistatin were shown in various diseases like arthritis, pneumonia etc.

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Our study is the first one that shows the anti-inflammatory effect of kalistatin in pulmonary hypertension. Kallistatin levels were low in pulmonary hypertension because of increased lung inflammation. Since no correlation between IL10 and TNF-alfa was found, probably they used different pathways in the pathogenesis of inflammation.

P1459 - MECHANICAL VENTRICULAR SUPPORT AS A BRIDGE TO HEART TRANSPLANT IN PEDIATRIC PATIENTS

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Introduction: Since July 2000 to November 2016, 119 pediatric patients where listed for cardiac transplant. Due the lack of suitable donors and the huge mortality rate during the waiting list we started with the use of mechanical cardiac support (MCS) as a bridge to transplant. This MCS, provides long time assistance in very young patients. Before using the MCS the mortality rate in this group was 50.%.
Material and Method: From March 2006, we assisted 44 patients. 43 with a Pulsatile Systems Berlin Heart (R) that consist in paracorporeal pumps and a pneumatic impulse consolé (Ikus(R),and one intrathoracic, with a continuos flow system (HeartWare®). Biventricular MSC 24 (55%); univentricular 20 (45%), when is usted by this way is necessary a strict monitoring to decide to assist or not the right ventricle. Diagnostics were: Dilated Cardiomyopathies 35 (80%), Restricted Cardiomyopathies 6 (14%) and Congenital Heart Disease 3 (6%). The continuos flow intrathoracic device (Heartware®) was used to provide left ventricular mechanical assistance in a 15 years old male patient, with high pulmonary pressure, transplanted after 10 months of support.
Results: The MCS time range was 3 to 954 days (X = 150). The age range of assisted patients was 10 to 192 months (X = 87). Survival on MCS: 67%, 27 were transplanted. One patient still remains on MCS, waiting for a suitable heart. Major complications: Neurological events due to Coagulopathie N = 10 (26,3%), Infections N = 4 (9%), Aortic pseudoaneurism N = 1 (2%). The mortality rate in waiting list drop off into a 20% (50/30%).
Conclusion: Mechanical Ventricular Support is an effective system to maintain alive the pediatric patients waiting a suitable heart donor. The artificial heart gives better results and improves condition of the pediatric patients before and after the transplant.

P1475 - MYCOTIC ANEURYSM AFTER BIVENTRICULAR ASSISTANCE DEVICE IN PATIENT SUBMITTED TO PEDIATRIC HEART TRANSPLANTATION

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Background: Aneurysm of the ascending aorta, particularly mycotic pseudoaneurym, is a very rare and potentially fatal complication in pediatric patients after heart transplantation, They are frequently associated with rupture and sepsis, with increased morbidity and mortality. Its management still remains a clinical and surgical challenge.
Objectives: To present the management of pediatric heart transplanted patient with mycotic aortic aneurysm after use of biventricular assistance device.
Case Presentation: A 8 year old girl was submitted to heart transplantation (HTX) in 2012 due to complex congenital heart disease. After four years of HTX, she presented with acute humoral rejection and cardiogenic shock, requiring circulatory assistance for 11 days, with complete recovery of cardiac function. After 6 months, she returned to our service with arterial hypertension, osteomyelitis in the right knee and superior vena cava syndrome. Echocardiogram showed a giant aortic aneurysm in ascending aorta. She was sent to the operating room and underwent an in situ graft replacement. Histological study revealed fungical infection. The postoperative course was uneventful and she was discharged after complete treatment of the mycotic infection.
Conclusion: Although the treatment of mycotic aortic aneurysm is difficult, surgical and clinical approach may present good results.

P1479 - CLINICAL OUTCOMES OF HEART TRANSPLANTATION IN A SMALL NORTHEAST CARDIAC PEDIATRIC CENTER IN BRAZIL

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Background: Heart transplantation (HTx) has become the standard care for children with end-stage heart failure refractory to medical or conventional surgical therapy, whether secondary to underlying congenital heart disease (CHD) or cardiomyopathy (CMP). Early and late mortality have significantly improved during recent decades in pediatric patients after HTx, although small center volumes are still associated with poorer outcomes. In Brazil, according to data of The Brazilian Registry of Transplants in 2014, pediatric HTx represents 11,25% of all heart transplants.
Objectives: To presente a 14-year experience of pediatric HTx of a northeast small referral center in Brazil, focusing on survival and long-term results.
Methods: All consecutive patients submitted to HTx from january 2002 to december 2016 were included in the study. Data collection consisted of demographic data, main indications and complications, immunosuppression therapy and mortality.
Results: There were 45 HTx in 43 patients during a period of 14 years. The two cases of re-transplants were due to graft dysfunction. Mean age at the time of HTx was 9,82 ± 5,58 years and the mean weight was 28,74 ± 15,92 kg. Twenty six (57,8%) patients were male. The indications of heart transplantation were cardiomyopathy (CMP) in 21 (46,7%) and CHD in 22 (48,9%) patients. The main complications were anemia (15,6%), systemic hypertension (11,1%) and hypercholesterolemia (11,1%). The main drugs for immunosuppression were cyclosporine (57,6%) and mycophenolate (60,6%). The mean follow up was 47,91 ± 34,81 months and the mean overall survival was 90,37 ± 8,35 months, without differences between patients with diagnosis of CMP or CHD.

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Conclusion: HTx is an acceptable therapeutic option for children and young adults, even in small centers with outcomes comparable to large centers.

P1533 - ACUTE CARDIOTOXICITY SECONDARY TO CHEMOTHERAPY TYPE SAINT JUDE ABOUT FOUR CASES

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Background: Among the forms of leukemia affecting children, acute lymphoblastic leukemia (ALL) is by far the most common. It is treated with chemotherapy, the most common, type Saint Jude. However, anthracycline cardiotoxicity remains the most risk to fear.
Objective: The aim of this study is to show the cardiotoxicity risk of chemotherapy type Saint Jude in children with acute leukemia.
Methods: This is a retrospective study of 04 cases with ALL, treated by chemotherapy type Saint Jude and complicated by cardiotoxicity. These patients were collected in clinical hematology department, Farhat Hached hospital-Sousse between 2011 and 2016.
Results: We collected four male children with a median age of 10 years [5 years -14 years], suffering from ALL. The diagnosis of acute leukemia was confirmed by myelogram. Immunophenotyping was in favor of a LLA-B in three children and of a LLA-T in a single patient. The karyotype objectified translocation (1.19) in a single patient. The pre-chemotherapy cardiac ultrasound was normal with a fraction of left ventricular ejection correct in all patients. These children were treated by chemotherapy type Saint Jude. The evolution was marked by the onset of respiratory symptoms such as orthopnea associated with a dry cough. Thus, the diagnosis of heart failure secondary to cardiotoxicity of anthracyclines was suspected and confirmed by echocardiography which objectified: LVEF altered (15-30%), a global hypokinesis with a left ventricular dilatation in all patients. Adequate medical treatment was instituted. Three children achieved complete remission of the acute leukemia with regression of respiratory symptoms and improving heart function. A child was died by his heart failure.
Conclusion: Anthracyclines (doxorubicin, daunorubicin. .) have largely proven effective in treating acute leukemia. However, they exert a dose-dependent, cumulative and often irreversible cardiotoxicity. Hence, the need for its early detection is the only effective way to improve the prognosis.

P1540 - PULMONARY HYPERTENION IN EXTREME PRETERM BORN INFANTS A SYSTEMATIC REVIEW AND META ANALYSIS

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Background/Hypothesis: Pulmonary Hypertension (PH) is known to complicate the course of extreme preterm born infants and is associated with poor outcomes in Bronchopulmonary Dysplasia (BPD). Prevalence, risk factors and outcome of PH are insufficiently known. Therefore our aim was to provide an up-to-date synthesis to examine these factors by performing a systematic review.
Materials/Methods: Databases Medline, EMBASE and The Cochrane Library were searched on the 12th of July 2016. Two authors reviewed titles/abstracts and full-text. Eligible studies reported one of the following: 1) PH prevalence, 2) patient characteristics of infants with and without PH, or 3) mortality. Studies without extreme preterm born infants were excluded. Thereafter data extraction and meta-analysis of prevalence, characteristics and mortality were performed. Heterogeneity was assessed using Cochran's Q-test and I^2 quantity.
Results: 1623 unique articles were identified, 22 articles were eligible for analysis. Meta-analysis, shown in table 1, assessed prevalence rates, risk factors and mortality. Prevalence of PH in severe BPD was 39% and infants with severe BPD were significantly more at risk for PH (Risk Ratio (RR) = 4.16). Infants with PH were also more at risk for mortality (RR = 4.17).
Conclusions: PH appears most prevalent in infants with severe BPD. However, due to heterogeneity between the eligible articles, prevalence rates for PH in infants with or without BPD could not be accurately estimated. Severe BPD appears a prominent risk factor for PH. Other risk factors for PH, were also risk factors for BPD. Available data did not allow for a distinction in risk factors between infants with and without BPD. Finally, PH increases risk of mortality. However, the effect of PH on mortality for infants without BPD remains unknown. In conclusion, current literature does not allow confirmatory conclusions on incidence, risk factors and outcome. Therefore, properly designed, prospective studies are needed.

Table 1. Results meta-analysis

Prevalence rate PH	**% (CI)**	***P-value***
No BPD	2 (0 - 8)	0.11
Mild BPD	4 (0 - 9)	0.01**
Moderate BPD	8 (2 - 16)	<0.001**
Severe BPD	39 (27 - 51)	<0.001**
Risk factors PH	**RR (CI)**	
Severe BPD	4.2 (2.9 - 6.0)	<0.001
SGA	2.1 (1.5 - 2.9)	<0.001
	SMD (CI)	
Low birth weight	-0.8 (-1.0 - -0.6)	<0.001
Low birth weight in BPD	-0.3 (-0.5 - -0.01)	0.060
Mortality	**RR (CI)**	
Infants with PH	4.2 (2.2 - 7.8)	<0.001

** p value < .01 for heterogeneity
Definition of abbreviations: RR = risk ratio; CI = 95% confidence interval; PH = pulmonary hypertension; BPD = bronchopulmonary dysplasia; SGA = small for gestational age; SMD = standardized mean difference.

P1547 - RISK FACTORS FOR LONG TERM ADVERSE EVENTS IN PEDIATRIC HEART TRANSPLANTATION IN SINGLE CENTER

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Introduction: Pediatric heart transplantation, the ultimate treatment option for end-stage heart failure has undergone major changes

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over the past three decades. However, some long-term complications are still left. We conducted this study to determine the relative risk factors for long-term adverse event associated with pediatric heart transplantation.
Method: The medical records of pediatric patients with heart transplantation in Asan Medical Center Children's Hospital, since 1997 were reviewed retrospectively. We classified long-term adverse events as death, re-transplantation, hemodynamically compromised rejection including cardiac allograft vasculopathy (CAV), persistent renal dysfunction, infection and post-transplant lymphoproliferative disease (PTLD). We assumed possible risk factors for major adverse events as age of transplantation, waiting time, primary diagnosis (cardiomyopathy or congenital heart disease), Severity of heart status(Korean Network for Organ Sharing system), CMV or EBV serology, type of immune suppressant and apply of mechanical circulatory support.
Result: For 62 eligible patients (36 males, median age at transplant is 10.9 months), overall mortality was 22.6%(n = 14; 10 year survival rate = 70.1%) and 5 patients underwent re-transplantation. Among these patients, 19 had rejection, 11 had infection, 7 had persistent renal dysfunction and 1 had PTLD. Nineteen patients (30.6%) had critical adverse event (CAE; death or re-transplantation) and the only risk factor for CAE is the type of immune suppressant; cyclosporin increase the risk of CAE than tacrolimus (p = 0.004, Odds ratio, 9.041; 95% CI 2.035-40.165). The other presumed-risk factors did not increased the prevalence of CAE significantly. In 6 patients who transplanted due to Fontan failure, only 1 died from graft failure. No other CAE was reported in this group.
Conclusion: Although the overall outcome of heart transplantation has been improved largely attributed to advance of immune suppressant, several risk factors, especially CAV is left and its evaluation in children has some limitations. The novel surveillance method suitable for pediatric transplant group should be studied.

P1560 - PEDIATRIC CARDIAC TRANSPLANTATION EXPERIENCE FROM INITIAL 30 CASES

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Introduction: Heart transplantation is the only definite therapy for children with end stage heart failure or those with irreparable complex single ventricle defects, not responding to optimal medical therapy. We present our experience with transplantation in patients <18 years of age.
Methods: 30 patients <18 yrs underwent cardiac transplantation between Jan 2012 and Nov 16. The Youngest was 11 months old. 24(80%) patients had idiopathic dilated cardiomyopathy, 3(10%) had restrictive cardiomyopathy, 3(10%) had previous palliation for single ventricle one of whom had only single lung perfused. 3 had familial cardiomyopathy, with 2 siblings receiving successful transplantation after bridging with assist device. 4(13.3%) patients were bridged with assist device prior to transplantation. Cardiac catheterisation was performed preoperatively and they were listed for tranplantation only if the PVRI was less than 5 woods units, with or without pulmonary vasodilators. All of them received protocolised immunosuppression with inductioin with basiliximab and maintanence with steroid, tacrolimus and mycophenolate. Donors age ranged from 12 to 40 years. 75% of the donors were outside the state, the mean ischemic time was 190 minutes. Custodial cardioplegia was used in all. 2 (6.6%) required post op ECMO support for RV failure of whom one survived.
Result: The mean duration of ventilation was 50 hrs and ICU stay was 8 days. There were 3(10%) early mortality (<30 days) and 2 (6.6%) late mortality after 6 months duration. 3 developed posterior reversible encephalopathy syndrome. Of whom one had early and one late death. All the three single ventricle patients including one with single lung had successful transplantation with excellent functional outcome.

P1561 - TRANSPLANTATION IN SINGLE VENTRICLE PATIENTS

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Heart transplantation as an option in the management of patients with single ventricle has long been in vogue. It can be introduced at any stage in the management of patients with single ventricle, at the first stage before any palliation or after Fontan failure. We present our experience in 3 young patients who received heart transplantation for single ventricle, one of them had heart transplantation into single lung. Patient 1 – 11 yr old girl with no previous palliation with, common AV canal, severe AV valve regurgitation, unbalanced ventricle with dysfunction, TAPVC supracardiac TAPVC with PS, underwent cardiac transplantation-her postoperative course was totally uneventful. Patient 2 – This 15 yr old boy had previous shunt and bilateral bidirectional glenn shunt, he had increased PVRI and ventricular dysfunction and was considered unsuitable for two ventricle repair or Fontan. He received cardiac transplantation and ligation of the left superior venacava as there he was discharged on day 30 after eventful postop course. Patient 3 – 18 yr old boy who had previous BT shunt and glenn. His right pulmonary artery was totally occluded and all forms of investigation failed to show any usable pulmonary artery segment. This was replaced by a leash of collaterals entering the right lung. He had heart transplantation to the single left lung after creating a PFO. He did well and was discharged on oxygen which was tapered and stopped after 40 days.
Conclusion: Transplantation into single lung is possible in patients with single ventricle. transplantation provided excellent palliation for single ventricle patients and can be considered in preference to high risk interim procedures.

P1575 - INSERTION OF LVAD STERNOTOMY AND THORACTOMY APPROACH – TECHNIQUE AND RESULTS

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Temporary mechanical support to a failing heart nonresponsive to inotropes is necessary either as a bridge to decision or bridge to transplantation. The available options are limited by their cost or lack of availability in small sizes. We present our technique which we believe is economical, easily reproducible and applicable in wide variety of situations. A sternotomy with limited pericardiotomy and thoracotomy approach is used to access the ascending aorta and the apex of the heart and also as a means for initiation of cardiopulmonary bypass if necessary. A conduit is sutured to both the areas, and a single lumen cannula is inserted into the apex, both are connected with appropriate connectors and

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after deairing is connected to centrifugal pump (Levitronix). The procedure was used in 8 patients with imminent decompensation due to end stage heart failure refractory to inotropes. One patient succumbed to intracranial bleeding. One patient died due to renal failure following heart transplantation. The youngest is 11 month 7 kg child awaiting transplantation. Adequate flows could be attained in all the patients. There was significant and in some cases dramatic improvement in end organ function following institution of LVAD. LVAD insertion using centrifugal pump and combined sternotomy, thoractomy approach is a useful lifesaving option for patients with endstage heart failure awaiting transplantation or as a bridge to decision.

P1592 - MYOCARDIAL FUNCTION ECHOCARDIOGRAPHIC EVALUATION IN HEART TRANSPLANTATION WITH LONGITUDINAL PEAK SYSTOLIC STRAIN

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Introduction: Acute cellular rejection is responsible for great morbidity after heart transplantation and its early detection is important in patient prognosis. The longitudinal peak systolic strain (LPSS) echocardiographic evaluation detects systolic dysfunction earlier than the commonly used ejection fraction measured by Teicholz method. There are few studies using the former method in pediatric heart-transplanted patients.
Objective: To evaluate the use of LPSS in early detection of myocardial dysfunction after heart transplantation in pediatric patients.
Methods: A retrospective study in pediatric heart-transplanted patients who underwent LPSS echocardiographic evaluation and myocardial biopsy within two months before or after echocardiography.
Results: In 22 patients evaluated, 12 (52.1%) had no acute cellular rejection in biopsy and 10 (43.5%) had mild cellular rejection (1 R). Mean LPSS values were -13.7% and -12.5% in patients with negative and positive biopsy results respectively. Student T test found no statistical significance between acute cellular rejection and LPSS value ($p = 0.508$).
Discussion: Previous studies have shown that heart-transplanted patients with no coronary artery disease have reduced LPSS values despite of normal global myocardial function. The present work showed reduced LPSS values in acute cell rejection, but also the same tendency among all patients, in accordance with literature. Other studies are necessary to find an association between LPSS values and myocardial dysfunction in pediatric heart transplantation.

Table.

Variable	Degree	n	Average	SD	Medium	Minimum	Maximum	P*
Strain	OR	12	-13,71	4,60	-15,10	-20,00	-5,30	0,508
	1R	11	-12,55	3,57	-12,50	-18,20	-6,60	
S LATVD	OR	12	7,67	2,15	7,00	5,00	11,00	0,114
	1R	11	6,36	1,57	7,00	3,00	8,00	
S SEPTO	OR	12	6,42	1,56	6,00	4,00	9,00	0,429
	1R	11	5,91	1,45	6,00	4,00	8,00	
S LATVE	OR	12	7,67	1,23	7,50	6,00	10,00	0,167
	1R	11	8,55	1,69	8,00	7,00	12,00	
S INF	OR	12	7,33	1,37	7,00	5,00	10,00	0,709
	1R	11	7,09	1,70	7,00	5,00	10,00	
S ANT	OR	12	6,92	1,24	7,00	5,00	9,00	0,431
	1R	11	7,64	2,69	7,00	4,00	13,00	

P1606 - POST TRANSPLANT LYMPHOPROLIFERATIVE DISEASE IS ASSOCIATED WITH STERNOTOMY DURING INFANCY AND LEFT VENTRICULAR HYPOPLASIA A POPULATION BASED RETROSPECTIVE REVIEW

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Background/Hypothesis: Pediatric heart transplantation has been available in Sweden since 1989; it is centralized to two centers, Gothenburg and Lund. As our unit in Gothenburg faced an increased rate of Post-transplant lymphoproliferative disease (PTLD), the objective of the study was to identify possible risk factors in our cohort of children.
Materials and Methods: This is a retrospective study of all 71 children who underwent heart transplantation (0–18 years of age) in Gothenburg, Sweden from 1989 to 2014.
Results: The overall incidence of PTLD was 15% (11/71). Mean age at transplantation was 9 years (0–17) and mean post-transplant follow-up time was 5,5 years (0.5–21.9) in the group that developed PTLD, compared with 10,2 years (0.02–25.2) for those who did not develop PTLD ($p = 0.062$). In our study group risk factors for PTLD were surgically palliated congenital heart defects ($p = 0.0003$), sternotomy during infancy ($p = < .0001$), hypoplastic left ventricle ($p = < .0001$), number of surgical events ($p = 0.0001$), mismatch concerning Epstein Barr virus infection ($p = 0.0001$), induction therapy with Antithymocyte globulin produced by Fresenius Medical Care (ATG-FRESENIUS) compared to Antithymocyte globulin produced by Genzyme (THYMOGLOBULINE) ($p = 0.028$) and immunosuppressive treatment with Tacrolimus compared with Ciklosporine ($p = 0.028$).
Conclusions: This study has three major findings. First, PTLD only developed in subjects born with congenital heart defects, especially univentricular defects with a hypoplastic left ventricle. Second, the vast majority (10/11) of subjects developing PTLD had undergone sternotomy as infants. Third, the number of surgical events carried a higher risk of PTLD.

P1610 - BOSENTAN FOR SEVERE PULMONARY HYPERTENSION ASSOCIATED WITH CONGENITAL HEART DISEASE IN CHILDREN A SINGLE CENTER OPEN LABEL PROSPECTIVE STUDY

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Background: Bosentan has been used in adult patients with pulmonary arterial hypertension(PAH). This open-label, prospective study was to evaluate the effectiveness and safety of Bosentan in children with severe PAH associated with congenital heart disease (PAH-CHD).
Methods and Results: Patients were enrolled from Department of Paediatric Cardiology of Beijing Anzhen Hospital and received Bosentan therapy for a minimum of 24 weeks. They were assessed by 6-minute walking distance (6MWD), transcutaneous oxygen saturation (SaO2), New York Heart Association functional class (NYHA-FC), haemodynamic parameters assessed by right- and left-heart catheterization and liver function before and after therapy. Of the 35 children diagnosed with severe PAH-CHD, mean

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age of (11.9 ± 5.0) years (1.5-18.9 years), 33 completed all follow ups (13 males and 20 females). After 24 weeks Bosentan treatment, the pulmonary vascular resistance index (PVRI) decreased from (16.9 ± 6.1) Wood units•m2 to (15 ± 5.6) Wood units•m2 (p = 0.012). In 28 children (>7 years), 6MWD increased from (452.5 ± 105.5) m to (502.8 ± 74.8) m (P = 0.003). NYHA-FC either improved or remained stable in all the patients. The mean SaO2 increased from (91.4 ± 3.6)% to (91.6 ± 4.1)% (P = 0.690), systemic vascular resistance (SVR) decreased from (20.4 ± 6.8) Wood units to (20.3 ± 4.9) Wood units•m2 (P = 0.966), and mean systolic PAP (PAPs) increased from (79.9 ± 11.0) mmHg to (83.7 ± 10.5) mmHg (P = 0.057). However, these differences were not statistically significant. All patients tolerated Bosentan therapy well with only one patient developed a transient increase in transaminase.
Conclusion: Bosentan has beneficial effects on haemodynamic parameters, exercise capacity, and is well tolerated by children diagnosed with severe PAH-CHD.

P1622 - IMPROVING RESULTS OF HEART TRANSPLANTATION FOR THE FAILING FONTAN

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Background/Hypothesis: The reported mortality of heart transplantation for patients with a failing Fontan circulation ranges from 10 to 50%. To determine if results have improved at our institution, we compared the outcomes of an early cohort (Group 1, 1990-2011, n = 18) with a recent cohort (Group 2, 2012-2016, n = 20).
Materials and Methods: Between 1990 and 2016, 238 patients underwent heart transplantation (OCT) at our institution. Of these, 38 patients had heart transplantation for a failing Fontan circulation. Sixteen patients (Group 1, n = 8; Group 2, n = 8) had a Fontan conversion or revision (with cardiopulmonary bypass) at a mean of 4.3 ± 4.2 years (median, 2.6 years) prior to OCT. Seven of these patients had their OCT within 18 months of the conversion/revision operation. Cross-sectional follow-up for survival status was obtained for all patients on October 16, 2016.
Results: Median age at OCT was 12.7 (Group 1) versus 10.9 years (Group 2) (p = 0.26*); mean age at OCT was 16.3 ± 12.4 (Group 1) versus 11.6 ± 7.4 years (Group 2) (p = 0.30, t-test). Median interval of Fontan to OCT was 7.3 (Group 1) versus 8.1 years (Group 2) (p = 0.91*). Operative mortality was 5 (28%, Group 1) versus 0 (Group 2) (p = 0.017*). *Wilcoxon rank sum test was used. Overall survival for the 38 Fontan patients was 87% at 1 year, 77% at 5 years, and 62% at 10 years.
Conclusions: In the recent era the results of heart transplantation for failed Fontan patients has improved markedly. These improved results should be considered in evaluating patients prior to Fontan conversion or revision attempts. Earlier referral of failing Fontan patients for OCT will improve patient outcomes.

P1653 - SYSTEMATIC LITERATURE REVIEW ON THE INCIDENCE AND PREVALENCE OF HEART FAILURE IN CHILDREN AND ADOLESCENTS

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Background: To assess the incidence and prevalence of heart failure (HF) in children and adolescents (age <18 years) over the last 20 years (1996-2016).
Methods: Cochrane Collaboration methodology was used to undertake this narrative systematic review. A search strategy was designed to retrieve studies in English from sources including Embase, Medline, recent conference proceedings, NHSEED, HTA, and WHO databases. Observational studies reporting incidence and/or prevalence of pediatric HF were included.
Results: 83 unique studies were included and categorized according to the method of reporting HF diagnosis: 1) primary diagnosis (n = 10); 2) HF associated with other cardiovascular diseases (CVDs) (n = 49); 3) HF associated with non-CVDs (n = 24). Most publications were from single-center studies. For primary HF, the incidence ranged from 0.87/100,000 (UK/Ireland) to 7.4/100,000 (Taiwan). A prevalence of 83.3/100,000 was reported in one large population-based study (Spain). HF etiology varied across regions. In lower income countries, lower respiratory tract infections and severe anemia were major causes, whilst in higher income countries, major causes of HF were cardiomyopathies and congenital heart disease. Key findings for categories 2 and 3 included a prevalence of HF associated with cardiomyopathies ranging from 36.1% (Japan) to 79% (US); associated with congenital heart disease from 8% (Norway) to 82.2% (Nigeria); associated with rheumatic heart diseases from 1.5% (Turkey) to 74% (Zimbabwe); associated with renal disorders from 3.8% (India) to 24.1% (Nigeria); and associated with HIV from 1% (US) to 29.3% (Brazil).
Conclusions: To our knowledge, this is the first systematic review of the incidence and prevalence of HF in children and adolescents, providing valuable insights into, and strengthening current knowledge of, pediatric HF. Although many studies were identified, heterogeneity in study design and diagnostic criteria limits the ability to compare regional data. There is a need to standardize definitions of pediatric HF to facilitate cross-regional data comparisons.

P1658 - THE ROLE OF CIRCULATING PROGENITOR CELLS AND CIRCULATING ENDOTHELIAL CELLS BEFORE AND AFTER CORRECTION OF CONGENITAL HEART DEFECTS

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Background: Circulating progenitor cells (CPC) and circulating endothelial cells (CEC) have been proposed as useful markers of severity and disease progression in patients with pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD). Limited information exists regarding CPC and CEC

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values after so-called definitive repair has been established. We sought to investigate these biomarkers in the setting of CHD with L-to-R shunts.

Methods: We are prospectively quantifying CPC and CEC before and after repair in a cohort of patients with CHD (n = 75). We report preliminary findings in 14 children (median age 4 years (2-12)) who have completed follow-up(FW). CPC (CD45dim,CD34 + , KDR2 +) and CEC (CD45-,CD31 + ,CD146 + ,CD105 +) were quantified in the SVC, pulmonary artery and femoral artery during catheterization prior and at least 6 months after repair. Values are expressed as median (range) or meanSD. Paired-t test was used accordingly. Statistical significance was accepted as p0.05.

Results: Defects included: VSD(n = 4); PDA(n = 7); ASD, AVSD and truncus arteriosus (n = 1 each). PDA and ASD patients underwent transcatheter device occlusion; the remainder underwent surgical repair. Median time for FW catheterization was 8 months(6-14). Table 1 shows hemodynamic variables and cellular counts before and after intervention; pulmonary artery pressure (PAP) normalized in all patients at FW. Most patients showed significant reduction of both CPC and CEC counts in all sample sites. However, in 4 children CEC counts increased, did not decrease and/or remained above reference value. In these patients, PAP at FW, although normal, was significantly higher than in the rest.

Conclusions: CPC and CEC levels decrease in patients with reversible PAH-CHD within few months after repair. This highlights the capacity of the pulmonary endothelium to mend once the hyperkinetic phase of cardiac L-to-R shunts is no longer present. Furthermore, it may guide appropriate management in patients that remain with elevated CEC counts, hence at risk of persistent or recurring PAH.

Table.

Variable	Before	After	p-value
Systolic PAP, mm He	53 ± 20	29 ± 3	.001
Mean PAP, mmHg	33 ± 15	15 ± 2	.001
PVRi, WU	2.9 + 0.97	2.5 ± 0.57	.325
PVRi/SVRi	0.23 ± 0.07	0.21 ± 0.05	.559
Qp:Qs	3.3 ± 0.69	1 ± 0.1	.001
CPC, cells/10^s			
SVC	51 ± 34	14 ± 12	.001
PA	53 ± 37	12 ± 11	.001
Systemic (FA/PV)	33 ± 27	12 ± 9	.019
CEC, cells/10^6			
SVC	72 ± 75	14 ± 11	.007
PA	70 ± 83	17 ± 25	.009
Systemic (FA/PV)	62 ± 74	14 ± 15	.023

Abbreviations: PAP pulmonary artery pressure; PVRi pulmonary vascular resistance index; PVRi/SVRi pulmonary vascular resistance index/systemic vascular resistance index ratio; Qp:Qs pulmonary to systemic flow ratio; CPC circulating progenitor cells; SVC superior vena cava; PA pulmonary artery; FA femoral artery; PV pulmonary vein; CEC circulating endothelial cells. Values are expressed as mean ± SD.

P1659 - CLINICAL PROFILE AND OUTCOME OF PERIPARTUM CARDIOMYOPATHY AMONG TEENAGER PATIENTS AT THE UNIVERSITY OF THE PHILIPPINES PHILIPPINE GENERAL HOSPITAL (UP PGH)

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Background: Peripartum Cardiomyopathy (PPCM) is a rare new-onset disease of the myocardium occurring in the peripartal period (within the last month of pregnancy to 5 months postpartum) resulting to decreased ventricular contractility (LVEF ≤ 45%) and heart failure. Multiparity, advanced maternal age, multifetal pregnancy, preeclampsia, gestational hypertension, and African-American race have been associated with peripartum cardiomyopathy. There is inadequacy of epidemiologic data of PPCM patients less than 19 years old. We determined the clinical profile and outcome of peripartum cardiomyopathy among teenagers admitted to our institution.

Materials and Methods: All 14-18 years old patients referred to the UP-PGH Section of Pediatric Cardiology from 2004-2013 diagnosed to have PPCM based on the criteria set by National Heart, Lung and Blood Institute (NHLBI) were included in the study. The demographic and clinical profile including age, parity, co-morbid conditions, and outcome; and 2D-Echocardiogram findings were extracted from the medical records. Mean and standard deviation were computed for continuous data, and frequency and percentages were presented for categorical data.

Results: Twenty five patients (mean age = 17years) diagnosed with PPCM were included in the study. Six patients (24%) had twin delivery. The most common co-morbidity was preeclampsia (10/25). Twenty-one developed symptoms of heart failure after birth. Most (24/25) presented in NYHA functional class III-IV. Majority had poor LV contractility (mean EF = 29%). Most (92%) had term deliveries, and 21 delivered vaginally. LV function improved in 18 mothers after 6 months. Four died from intractable heart failure.

Conclusion: PPCM occurred not infrequently among 14-18 year old patients. Pre-eclampsia is the most common co-morbidity which is consistent with other studies. More in-depth study on risk factors of PPCM in this particular population is warranted.

Table 1. Demographic and clinical profile of 25 patients with peripartum cardiomyopathy

Age, mean + /- sd (range)	17 years old + /- 1.3 (14-18 years old)
Parity, n (%)	
1	21 (84%)
2	2 (8%)
3	2 (8%)
4	0
Age of Gestation, mean + /- sd (range)	37 wks + /- 0.8 (35 wks – 39 wks)
Number of Sex Partners (%)	1 sexual partner (84%) 2 sexual parters (8%) 3 or more sexual partners (8%)
Type of Birth, n (%)	
Single	19 (76%)
Twin	6 (24%)
Triplet	0
Onset of Symptoms, n (%)	
After delivery	23 (92%)
Before delivery	2 (8%)
NYHA Functional class, n (%)	
I	1 (4%)
II	0
III	18 (72%)
IV	6 (24%)
Mode of Delivery, n (%)	
SVD	21 (84%)
CS	3 (12%)
OFE	1 (4%)
Birth Weight, n (%)	
AGA	24 (96%)
SGA	1 (4%)
LGA	0

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Table 2. Echocardiographic parameters of 25 patients with peripartum cardiomyopathy

Ejection fraction, mean + /- sd (range)	29 % + /- 11.9 (4-50%)
Fractional Shortening, mean + /- sd (range)	12.7 % + /- 5.5 (2-20%)
LVEDD mean + /- sd (range)	5.3 cm + /- 12.8 (3.5-7.9 cm)
LVESD, mean + /- sd (range)	4.7 cm + /- 7.2 (3.6-6.4 cm)
Valvular Regurgitation, n (%)	
Mitral valve	23(92%)
Tricuspid valve	15(60%)
Aortic valve	9(36%)
Chamber Enlargement, n (%)	25(100%)

Table 3. Outcome of 25 patients diagnosed to have PPCM

Discharged, n (%)	18 (72%)
Mortality, n (%)	4 (16%)
Lost to Follow up, n (%)	3 (12%)

P1670 - HEART TRANSPLANTATION IN PEDIATRIC AND ADULT WITH CONGENITAL HEART DISEASE CURRENT STATUS

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Introduction: Cardiac transplantation is considered the therapeutic treatment for refractory heart failure. Endomyocardial biopsy (EMB) is the gold standard for diagnosis, allowing adequate treatment in cases of graft rejection.
Materials and Methods: Retrospective study of patients submitted to EMB from January 1, 2015 to December 31, 2015, at the Heart Institute (HCFMUSP). The objective of the study was to correlate the EMB with the time of transplantation, clinical and echocardiographic findings.
Results: A total of 117 biopsies were evaluated in 51 patients in this period. 56 (47.8%) did not present cell rejection (0 R), 36 (30.7%) presented mild rejection (1 R), and in 18 biopsies (15.3%) there was moderate (2 R) rejection. Of the 51 patients who underwent biopsy, 17 (33.3%) were in the first year after transplantation. 5 (9.8%) of the patients were in the immediate postoperative period. Rejection 2 R occurred in 13 cases, and in 5 (38.4%) more than one episode occurred. One of the patient (7.6%) died. Of these 13 patients, 10 (76.9%) used tacrolimus, 3 (23.1%) of cyclosporine and all cases were taking mycophenolate sodium. We also observed that of the 13 cases with cell rejection (IIR), 8 (61.5%) presented echocardiographic changes. These were ventricular hypertrophy in 62.5% of the cases, ventricular diastolic dysfunction in 37.5%, ventricular systolic dysfunction in 25% and worsening of atrioventricular valve regurgitation in 25% of cases. There was no complication after the EMB procedure.
Conclusions: EMB is a low risk procedure that allows the diagnosis and treatment of pediatric and adult patients undergoing cardiac transplantation. The echocardiogram is a noninvasive method that has shown promise tool for rejection diagnosis.

P1714 - PEDIATRIC HEART TRANSPLANTATIONS IN A NATIONAL PEDIATRIC HOSPITAL IN ARGENTINA

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Background: Heart transplantation is the most appropriate surgical option for terminal heart failure in childhood. The lack of suitable donors is the biggest problem to develop Pediatric Cardiac Transplant Program.
Materials and Methods: This is a prospective, retrospective, transversal and observational paper. From July 2000, 119 patients have been listed for heart transplantation. Age: 1 to 230 months; Female: 52% ; Male: 48%. Range in waiting list: 1 to 3351 days. Diagnosis: Dilated Cardiomyopathy 85; Congenital Heart Disease 18; Restrictive Cardiomyopathy 13; Pulmonary Hypertension 2 ; Retransplant 1.
Results: Three Patients (16, 32, 7, months old) were transplanted with an ABO incompatible heart donor using the "West Protocol". Due to the lack of suitable donors and its consequence of high mortality on waiting list, in 2006 we have started to use Mechanical Assistance to let patients, in terminal condition, survive and have a chance for transplantation. Fifty-four heart transplants where done in 53 patients The actuarial survival rate in waiting list showed a huge improvement with the use of the Mechanical Cardiac Support (MCS); 26 patients were assisted with the MCS before transplantation. Age: 13 to 198 months; weight: 6 to 90,0 kg.; univentricular MCS: 14 (54%) and biventricular MCS: 12 (46%). No surgical deaths and no REDO because surgical bleeding in the MSC series. Five patients where assisted with ECMO after transplantation, due to primary heart failure; with good results in all of them after ECMO weanning. The triple scheme treatment for immunosuppression was: azathioprine, cyclosporine and steroids.
Conclusions: A Cardiac Transplant Program in a Pediatric Public Hospital; in Argentina; provides the opportunity for heart transplantation to infants and pediatric population with terminal heart failure.

P1735 - ECHOCARDIOGRAPHIC ASSESSMENT OF INFANTS WITH CONGENITAL DIAPHRAGMATIC HERNIA AND ASSOCIATED PULMONARY HYPERTENSION

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Introduction: Congenital diaphragmatic hernia (CDH) remains a challenging neonatal condition with a number of sequelae. In particular there is a lack of published evidence documenting the long term effects and outcomes of pulmonary hypertension (PH) in repaired CDH patients.
Aim: The aim was to document the long term echocardiographic assessment of patients with repaired CDH, in particular those showing evidence of post-operative PH.
Method: Retrospective analysis of infants with CDH treated at the Royal Hospital for Children, Glasgow between February 2009 and February 2015. The pre discharge; first discharge and most recent echocardiogram were identified. Parameters were identified that would delineate evidence of PH and assess left/right ventricular

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function. These included tricuspid regurgitation (TR); right ventricular size; systolic and diastolic functional assessment, including LV global speckle strain imaging and tissue Doppler imaging.
Results: Data was available for 62 patients of whom 47 (75.8%) survived to discharge. Median number of scans performed during hospital admission was 4.3 (IQ range 1-3), with final scans performed a median of 19 days before discharge (IQ range 3-24). 11 (17.5%) children were discharged on oral sildenafil, none were discharged on Bosentan therapy. The most common echocardiographic measurements assessed were the presence and velocity of TR, fractional shortening and ejection fraction, (82.3%, 75.8%, 66.1%, and 66.1% respectively). TR was present in 27 patients (43.5%), with a mean Vmax of 2.0 m/s (IQ range 0-3.2). Following discharge, 13 patients (21.0%), underwent an echocardiogram; median 117 days post discharge (IQ range 20-181).
Conclusion: There is currently no standardised approach for echocardiographic assessment of CDH patients. Our findings support the development of a standardised protocol for long term echocardiographic assessment of patients with repaired CDH.

P1744 - PULMONARY HYPERTENSION AMONG 5 TO 18 YEAR OLD CHILDREN WITH SICKLE CELL ANEMIA IN NIGERIA

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Background: Pulmonary Hypertension (PHT) is a significant cause of mortality in patients with sickle cell anemia (SCA). Few studies on PHT in SCA have been carried out in children. This study aimed to estimate the prevalence of PHT in children with SCA and determine its clinical and laboratory correlates.
Materials and Methods: In this cross sectional study, evaluation involved obtaining bio-data, history and physical examination findings in 175 SCA subjects with genotype SS aged 5 to 18 years and 175 age and sex matched controls with genotype AA. PHT was determined using echocardiography derived peak Tricuspid Regurgitant Velocity (TRV) > / = 2.5 m/s as a marker. Complete blood count (CBC), lactate dehydrogenase (LDH) assay, reticulocyte count, fetal hemoglobin estimation as well as Human Immunodeficiency Virus (HIV) I and II, Hepatitis B Virus (HBV) and Hepatitis C Virus (HCV) screening were done for patients with SCA.
Results: The mean peak TRV of subjects with SCA and controls was 2.2 ± 0.4 m/s and 1.9 ± 0.3 m/s respectively and prevalence of PHT among children with SCA and controls was 22.9% and 2.3% respectively. PHT in SCA correlated negatively with Body Mass Index, hematocrit and hemoglobin. A loud second heart sound was independently associated with PHT.
Conclusion: This study affirms that PHT prevalence is high in children with SCA and a loud second heart sound is associated with its occurrence.

P1753 - THE ROLE OF ACE2 ANG (1 7) MAS PATHWAY ON CARDIAC REMODELING IN PRESSURE OVERLOADED LEFT VENTRICLE MODEL

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Background: In the pressure overloaded left ventricle, left ventricular remodeling and retrograde pulmonary hypertension (PH) will develop. Subsequently, PH will lead to right ventricular remodeling and failure. And, the decreased cardiac expressions of ACE2-Ang-(1-7)-Mas related proteins in both ventricles are hypothesized simultaneously. The activator of ACE2 will attenuate the development of cardiac remodeling.
Methods: In ascending aora-banded (AOB) Wistar rat for six weeks, the hemodynamic measurements including left atrial pressure and pulmonary arterial pressure are arranged in the AOB rats and sham-operated rats. The cardiac expressions of ACE2, Ang-(1-7) and Mas will be checked.
Results: There were pressure-overloaded left ventricular hypertrophy, interstitial fibrosis and perivascular fibrosis, increased left atrial pressure, and increased pulmonary arterial pressure noted in the aortic banded rats. In addition, the cardiac expression of ACE2, Ang-(1-7) and Mas in both ventricles were decreased simultaneously. Very importantly, DIZE, ACE2 activator, could exert preventive and therapeutic effects on pathological findings such as hypertrophy and fibrosis, and hemodynamics such as left atrial pressure and pulmonary arterial pressure.
Conclusions: The ACE2/ Ang-(1-7)/Mas pathway could play an important role of pathophysiology of hypertrophic ventricle, and their pharmacologic modification exert the beneficial effects on the development of cardiac hypertrophy especially in pulmonary hypertension.

P1756 - METABOLIC REMODELLING IN RIGHT VENTRICULAR ADAPTATION TO PRESSURE LOAD

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Background: Because of improved techniques, more patients with congenital heart diseases are exposed to right ventricular (RV) chronic pressure load (PL) which makes them prone to develop RV failure. Disturbed cardiac metabolism, observed in end-stage RV failure, has been hypothesized as mechanism of deterioration. Most animal models of RV PL are focused on end-stage RV failure. However, to stop pathological processes contributing to failure, early intervention is necessary. We aimed to characterize time dependent remodelling and metabolic changes in a rat model of RV PL.
Material and Methods: Male Wistar WU rats (160-180 gram) were subjected to pulmonary artery banding (PAB, n = 25) or sham surgery (n = 13). RV adaptation to pressure load, including remodelling and metabolism were assessed at three different time points (two, five and twelve weeks). We assessed mitochondrial respiratory capacity with fatty acids (FA) or glucose as substrates (Oroboros). RV function was assessed by clinical score and echocardiography. RV remodelling was assessed by weights, echocardiography, histology and qPCR.
Results: PAB induced significant RV PL, reflected in RV hypertrophy and fibrosis, reduced RV function, septal bulging and upregulation of mRNA levels NPPA (see table for data). All animals showed preserved cardiac output, congruent with absence of overt clinical failure. At early adaptation to PL (two weeks), mitochondrial

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respiratory capacity for both glucose and FA tended to decrease. During prolonged PL (five and twelve weeks) the ratio of mitochondrial respiratory capacity of glucose vs. FA increased. At twelve weeks, glucose respiration was significantly increased, suggesting compensatory mechanisms in glucose metabolism.
Conclusion: This model of chronic RV PL suggests an early dysregulation of mitochondrial respiration with respiratory compensation overtime build on glucose metabolism. These data may help target therapeutic metabolic remodelling in the chronically PL RV.

Table 1.

	2 weeks		5 weeks	
	SHAM (n = 4)	**PL (n = 5)**	**SHAM (n = 4)**	**PL (n = 11)**
Weights				
Body Weight (g)	280 ± 15	254 ± 22	341 ± 13	322 ± 28
Right Ventricle (g)	0.17 ± 0.02	0.36 ± 0.06*	0.18 ± 0.01	0.48 ± 0.05*
(RV/Tibia Length)-100	4.8 ± 0.4	10.8 ± 1.2*	4.8 ± 0.4	12.8 ± 1.4*
Echocardiography				
LV CO (ml/min)	139 ± 15	129 ± 15	127 ± 21	123 ± 26
PAB gradient (mmHg)	5.3 ± 0.8	53.4 ± 9.7*	4.4 ± 0.2	77.9 ± 18.8*
TAPSE (mm)	2.8 ± 0.5	2.4 ± 0.2	2.8 ± 0.2	2.0 ± 0.4*
EI End Systolic	1.2 ± 0.2	0.6 ± 0.2*	1.2 ± 0.1	0.6 ± 01*
EI End Diastolic	1.1 ± 0.1	0.7 ± 0.2*	1.0 ± 0.0	0.7 ± 0.1*
qPCR				
NPPA (Fold Change)	1.0 ± 1.2	43.3 ± 16.1*	0.4 ± 0.0	39.9 ± 17.5*
MYH 7/6 (Fold Change)	1.0 ± 1.2	13.6 ± 5.5*	2.5 ± 0.8	18.1 ± 7.9*
Immunohistochemistry				
RV CCSA (um^2)	376.4 ± 31.9	746.8 ± 152.0*	380.0 ± 133.3	1076.5 ± 161.0*#
RV Masson Trichrome (%)	2.8 ± 1.2	7.1 ± 4.4	1.4 ± 0.4	9.9 ± 3.7*
Mito. Max Respiration				
Pyruvate O_2 Consumption (nmol/min/mg)	652.2 ± 145	416.5 ± 107	528.7 ± 89	584.1 ± 0.45
Palm. CoA O_2 Consumption (nmol/min/mg)	285.7 ± 109	186.5 ± 55.7	320.86 ± 72.5	233.15 ± 71.9
Pyruvate/Palm.CoA Ratio	2.46 ± 0.45	2.47 ± 0.99	1.70 ± 0.34	2.48 ± 0.37*
	12 weeks			
	SHAM (n = 5)	**PL (n = 9)**		
Weights				
Body Weight (g)	399 ± 24	393 ± 64		
Right Ventricle (g)	0.19 ± 0.03	0.50 ± 0.09*		
(RV/Tibia Length)-100	4.8 ± 0.4	12.3 ± 2.1*		
Echocardiography				
LV CO (ml/min)	136 ± 15	158 ± 39		
PAB gradient (mmHg)	4.3 ± 0.4	82.2 ± 13.4*		
TAPSE (mm)	2.76 ± 0.3	2.21 ± 0.2*		
EI End Systolic	1.0 ± 0.2	0.7 ± 0.0*		
EI End Diastolic	1.0 ± 0.1	0.7 ± 0.1*		
qPCR				
NPPA (Fold Change)	3.5 ± 6.0	28.5 ± 14.1*		
MYH 7/6 (Fold Change)	4.2 ± 5.9	17.4 ± 7.3*		
Immunohistochemistry				
RV CCSA (um^2)	407.8 ± 73.7	1221.6 ± 238.0*#		
RV Masson Trichrome (%)	3.7 ± 2.0	7.3 ± 3.5		
Mito. Max Respiration				
Pyruvate O_2 Consumption (nmol/min/mg)	532.7 ± 40.56	677.2 ± 144*		
Palm. CoA O_2 Consumption (nmol/min/mg)	284.9 ± 36.5	269.4 ± 66.7		
Pyruvate/Palm.CoA Ratio	1.70 ± 0.13	2.56 ± 0.48*		

Supplemental table. PL = pressure load. RV = right ventricle. LV CO = left ventricle cardiac output. PAB = Pulmonary Artery Banding. TAPSE = tricuspid annular plane systolic excursion. EI = eccentricity index. CCSA = cardiomyocyte cross-sectional area. Mito. Max Respiration = State Uncoupled. Values are MEAN ± SD, * = < 0.05 vs. SHAM (Students t-test). # = < 0.05 vs. PL 2 weeks (ANOVA with Bonferroni post hoc correction).

P1773 - CURRENT ERA OUTCOMES OF POST TRICUSPID SYSTEMIC TO PULMONARY SHUNT WITH PULMONARY HYPERTENSION

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Background: Simple post-tricuspid systemic to pulmonary shunt such as ventricular septal defect (VSD), patent ductus arteriosus (PDA) and aortopulmonary window (APW) is common. Untreated significant shunt subsequently leads pulmonary arterial hypertension (PAH).
Objective: To define survival and prognosis of patients with isolated post-tricuspid systemic to pulmonary shunt and PAH. *Methods:* Data of consecutive patients with hemodynamic confirmation by cardiac catheterization of PAH due to isolated post-tricuspid systemic to pulmonary shunt in 1999 - 2015 were collected. Clinical outcomes and survival were recorded. Risks of mortality and functional class (Fc) worsening were performed.
Results: At a median follow-up of 4.6 years (1-16.1 years), 116 patients having PAH associated with VSD (n = 78), PDA (n = 37) and APW (n = 2) were reviewed. The age at the diagnosis was 19.1 ± 17.1 years. The mPAP measured 69.9 ± 17.5 mmHg and baseline PVR was 19.6 ± 16.4 WU•m2. 26 patients were initially defined as Eisenmenger syndrome. 84 patients (72.4%) underwent defect closure and 20 had persisted PAH after correction (22.8%). Overall, worsening Fc and mortality was 6.9% and 2.6%. Risk of adverse events was final PVR >8 WU•m2 (p = 0.01) and elevated right atrial pressure (p 0.04). Survival at 10 and 15 years was entirely 100% in patients with PVR <8 WU•m2 while it was 93.4% and 90.3% in patients with PVR >8 WU•m2 (p = 0.01) regarding of anti-pulmonary hypertensive drugs.
Conclusion: PAH associated with isolated post-tricuspid systemic to pulmonary shunt has a high survival rate in contemporary era. PVR modulates the strategies of treatment and outcomes. Aggressive medical therapy should be considered in patients with PVR >8 WU•m2 after pulmonary vasodilator testing.

P1777 - ANGIOTENSIN CONVERTING ENZYME INHIBITORS MAY CAUSE THE NON OSMOTIC RELEASE OF PLASMA ARGININE VASOPRESSIN

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Background: Angiotensin-converting enzyme inhibitors (ACEI's) are an important medication in the treatment of congestive heart failure. However, ACEI's may cause harmful side effects, such as the non-osmotic control of plasma arginine vasopressin (pAVP) secretion, leading to the syndrome of inappropriate secretion of antidiuretic hormone (SIADH).
Materials and Methods: Thirty-two pediatric patients who received ACEI between January 2015 and November 2016, were enrolled in this study. The laboratory parameters were measured before and after ACEI administration. Among 10 patients with the osmotic regulation of pAVP release before ACEI administration, we compared pAVP levels between before and after ACEI administration. The non-osmotic release of pAVP was defined that pAVP levels were not suppressed when plasma osmolality (pOsm) was below the osmotic threshold for pAVP secretion. Patients were diagnosed as SIADH with the diagnostic criteria; serum sodium concentration (sNa) <135 mEq/L, pOsm <280 mOsm/kg H2O, urinary osmolality >100 mOsm/kg H2O, and urinary sodium concentration >20 mEq/L.
Results: The mean age was 4.0 ± 5.3 years. Seven patients received cilazapril, and three patients received enalapril. There were no differences between before and after receiving ACEI in pOsm (284.4 ± 7.1 vs. 283.5 ± 7.3 mOsm/kg H2O; p = 0.81), sNa concentration (138.1 ± 3.8 vs. 137.6 ± 2.9 mEq/L; p = 0.78), and pAVP levels (2.0 ± 1.2 vs. 2.6 ± 2.2 pg/mL; p = 0.38). Four patients (40.0%) had the non-osmotic release of pAVP, and one of them was diagnosed as SIADH without symptoms.

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Conclusions: This study suggested that ACEI could cause the non-osmotic secretion of pAVP.

P1830 - CARDIAC TRANSPLANTATION IN SURVIVORS FOLLOWING TREATMENT FOR PEDIATRIC MALIGNANCY

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Background/Hypothesis: Outcomes from malignancy occurring during infancy and childhood are improving. However, secondary side-effects can compromise quality of life and survival, including cardiac failure from chemotherapy/radiation. The timing of when cardiac transplant is performed, how patients are supported after listing but before transplant and the transplant outcomes in cancer patients are unknown. This data could inform use of temporary/destination extracorporeal support devices and impact the practice of cardiac transplant following malignancy diagnosis.
Materials & Methods: A retrospective analysis of patients ≤25 years old receiving cardiac transplants in the United States following their malignancy diagnosis was undertaken. The comprehensive United Network for Organ Sharing (UNOS) database from 1988-2015 was queried and abstracted. Patients were analyzed by: age ≤25 or >25 years old; pre-transplant malignancy diagnosis; type of any extracorporeal support pre-transplant; post-transplant recurrent malignancy; survival at censoring of database, among other attributes explored.
Results: From complete records 1988-2014, 59,705 total cardiac transplants were performed in the United States. Pre-transplant malignancy occurred in 2,265 patients, 2064 adult and 201 pediatric. The most common pediatric malignancy diagnoses included 55 leukemia/lymphoma and 37 sarcoma patients; 77 were "unknown". 105 pediatric patients were supported with extracorporeal devices pre-transplant including extracorporeal membrane oxygenation (ECMO) and multiple varieties of ventricular assist devices (VAD). There were 340 recurrent malignancies post-transplant occurring in 291 adult and pediatric patients; only 13 recurrences occurred in 9 pediatric patients.
Conclusions: Cardiac transplant for cardiac failure from chemotherapy/radiation can result in positive outcomes for pediatric cancer patients. However, not all potential transplant candidates may have been offered the option and thus are not included in UNOS database. Further systematic analysis of all patients' courses, perhaps linking UNOS to other registries, could optimize candidate selection and define potential role of long-term or destination VAD support in their management.

P1890 - DIAGNOSTIC CRITERIA OF HEMODYNAMICALLY SIGNIFICANT PATENT DUCTUS ARTERIOSUS IN PRETERM NEWBORNS

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Objective: To improve the accuracy of diagnosis of hemodynamically significant ductus arteriosus (HSDA) in preterm infants by determining objective clinical and Doppler echocardiographic criteria. The study involved 93 newborns of 24-35 weeks gestation (WG). Doppler echocardiography was conducted in the first days of life on a daily basis. The newborns were grouped into: Group 1 (n = 26) with weight 779.5 ± 63.4 g of 24-29 WG, Group 2 (n = 29) – weight 1297.1 ± 112.6 g of 30-34 WG, Group 3 (n = 38) with weight 1859.1 ± 118.1 g of 32-35 WG.
Results: Doppler echocardiography showed HSDA in 13 (50.0%) ($p < 0.05$) Group 1 children in accordance with the criteria elaborated by A.Sehgal, P.J.McNamara (2009); LV dilatation with hypertrophy of the wall and interventricular septum in 77.4% (p1\2; 1/3 < 0.05), RV dilatation in 82.8% (p1\2; 1/3 < 0.05), LA dilatation in 100% (p1\2; 1/3 < 0.05), 1st-2nd degree regurgitation on the tricuspid and pulmonary valves in 65.6% (p1\2; 1/3 < 0.05), increase in the average pressure in the pulmonary artery in 78.5% (p1\2; 1/3 < 0.05), disturbance of ventricular diastolic function by slow relaxation in 100% (p1\2; 1/3 < 0,05) of children. We identified additional Doppler echocardiography data in neonates who were rendered respiratory care with FiO2 40% or more, particularly the probability of false-negative result due to temporary functional closure of PDA. Furthermore, lack of reverse flow in the anterior cerebral artery and/or middle cerebral artery.
Conclusion: Morphological changes in cardiac chambers and clinical deterioration of newborns' state demonstrate the need to address the issue of hemodynamic significance ductus arteriosus. It is important to focus on the morphological changes in heart chambers, clinical condition of the patient and oxygen dependence.

P1955 - IMPLEMENTATION OF A CLINICAL PRACTICE GUIDELINE FOR GENERAL ANAESTHESIA OR PROCEDURAL SEDATION FOR CHILDREN WITH IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION

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Background: Children with idiopathic pulmonary arterial hypertension (IPAH) are a high-risk group who require diagnostic and invasive procedures with general anaesthesia (GA) or procedural sedation (PS). Complications including death, are reported more frequently in this population, but the true risk has not been quantified.
Aim: Review clinical outcomes, considering major complications, mortality and impact of a clinical practice guideline (CPG) for IPAH patients from a single paediatric tertiary centre (RCH, Melbourne).
Methods: A retrospective review using medical records and institutional databases was undertaken on patients with IPAH who underwent GA or PS as part of the state-wide pulmonary hypertension (PH) service. Patients were stratified into 3 groups: before availability of PH specific therapies (1.1.1980-31.12.1999), after (1.1.2000-31.12.2012) and following implementation of an institutional CPG (1.1.2013-1.12.2016). Data collected included disease severity, details of clinical management, and major complications occurring within 30 days of GA or PS. Major complications were defined as death, unplanned intensive care, cardio-pulmonary resuscitation, ECMO, intubation and pulmonary hypertensive crisis. Standard statistical methods were applied.
Results: During the study period, 41 patients underwent 107 procedures including 17 performed in the era without specific PH therapy, 62 in the era of available PH specific therapy and 28 utilizing the new CPG. Death occurred in 12% of untreated cases

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and 5% of treated era patients (p = 0.16). There were no deaths post CPG implementation. Major complications were significantly greater in the untreated era, representing 41% of cases, reducing to 6% and 11% respectively in the treated era and post CPG era (p = 0.002).

Conclusions: Mortality and morbidity in the form of major complications has markedly reduced over time as interventions undertaken in children with advanced disease increased. Implementation of a specific CPG for this vulnerable group with IPAH has promoted institutional awareness, education and overall improved care.

Table.

Descriptive Variable	Untreated era patients (1980-1999)	Treated era patients (2000-2012)	Treated era patients post CPG (2013-2016)
Patient numbers	11	17	13
Median age at IPAH Diagnosis (years)	10.3 (4.2, 11.7)	6.9 (4.8, 11.2)	7.3 (4.2, 11.3)
Median age at Time of Intervention (TOI)	9.5 (6.6,11.9)	11.2 (7.3, 14.3)	10.4 (5.0, 16.4)
Median duration of follow-up (years)	0.65 (0.1, 4.9)	5.14 (3.1, 9.5)	5.2 (2.7, 7.9)
Interventions with GA or PS	17	62	28
	GA = 16	GA = 50	GA = 20
	PS = 1	PS = 12	PS = 8
Types of Interventions	RHC = 11	RHC = 32	RHC = 7
Right heart catheter (RHC)	CVAD = 0	CVAD = 23	CVAD = 13
Central venous access device (CVAD)	Other = 6	Hybrid = 3	Hybrid = 1
		Other = 4	Other = 7
Gender distribution	Female = 3 (27%)	Female = 11 (63%)	Female = 7 (53%)
WHO class at TOI	I = 1 II = 6	I = 2 II = 22	I = 2 II = 5
	III = 9 IV = 1	III = 34 IV = 4	III = 11 IV = 10
	59% class III or IV	61% class III or IV	75% class III or IV
PAH status at TOI	Newly diag /untreated =	New/untreated = 16	Newly diag/ untreated = 3
	17	Stable on PH therapy = 31	Stable on PH therapy = 13
		Unstable on therapy = 15	Unstable on therapy = 12
Echocardiogram RVH & RV dilatation at TOI	Unavailable data	RVH & RV dilatation = 36 cases (58%)	RVH & RV dilatation = 20 (80%)
Baseline mean PAP (mmHg)	48.8	47.7	64.3
Major complications	7/17 cases = 41% **Mortality @ 30 days** = 2 cases both under GA = **12%**	4/62 cases = 6.4% **Mortality @ 30 days** = 3 cases, all with GA = **4.8%**	3/28 cases = 11% **Mortality @ 30 days = 0 cases, 0%**

P1975 - RIGHT VENTRICULAR VASCULAR COUPLING RATIO IN PEDIATRIC PULMONARY ARTERIAL HYPERTENSION COMPARISON OF INVASIVE AND NONINVASIVE MEASUREMENTS

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Background/Hypothesis: In pediatric pulmonary arterial hypertension (PAH), right ventricular (RV) failure is the main cause of death. RV-vascular coupling ratio (VVCR) is gaining interest as it describes contractile response to afterload. Conventional measurement of VVCR by cardiac catheterization (VVCRs) is invasive with associated risks. Therefore, noninvasively estimated VVCR (VVCRm) using cardiac magnetic resonance (CMR) may offer advantages in children with PAH. We aimed to compare VVCRm to VVCRs in pediatric PAH and tested two hypotheses: 1) VVCRm corresponds with VVCRs; and 2) both represent disease severity.

Methods: Patients diagnosed with idiopathic and associated PAH without post-tricuspid shunt, who underwent catheterization and CMR within 90 days at two specialized centers for pediatric PAH were retrospectively studied. VVCRs by single-beat pressure method was compared with VVCRm defined as stroke volume/end-systolic volume ratio, using correlation, regression analysis and Bland-Altman plots. Both were correlated with clinical measures of disease severity (World Health Organization functional class [WHO-FC], pulmonary vascular resistance index [PVRi], mean pulmonary artery pressure [mPAP], mean right atrial pressure [mRAP], RV cardiac index [RV CI]).

Results: Twenty-seven patients were included with median age at diagnosis 9 years (IQR: 3,7–14) and at enrollment 14 years (IQR: 11–17), PVRi 7,6 WU × m2 (IQR: 4,1–12,0) and median mPAP 40 mmHg (IQR: 28–55) (table 1). VVCRs and VVCRm showed a strong correlation (r = 0,81, p < 0,001) with a mean difference of 0,2 and 95% of the differences between -0,2 and 0,6. Both correlated with WHO-FC and hemodynamics (table 2).

Conclusion: Non-invasively measured VVCRm is feasible in pediatric PAH and comparable to invasively assessed VVCRs. Both VVCRm and VVCRs correlate with functional and hemodynamic measures of disease severity. The role of VVCR in predicting outcome and its use as treatment goal in pediatric PAH requires further clinical investigation.

Table 1. Patient characteristics

	N	Value (percentage) or median (IQR)
Age at PAH diagnosis (years)	27	9,0 (3,7- 14,0)
Female	27	16 (59)
Etiology	27	
IPAH/HPAH		18 (67)
APAH-CHD, repaired		5 (19)
APAH-other		4 (15)
WHO	22	
I-II		13 (59)
III – IV		9 (41)
PVRi (WU x m^2)	27	7,6 (4,1-12)
mPAP (mmHg)	27	40 (28 - 55)
mRAP (mmHg)	27	6,0 (4,0 - 8,0)
RV CI (L/min/m^3)	27	4,1 (3,2-4,5)

Table 2. $VVCR_m$, $VVCR_s$, and disease severity

		VVCRm		VVCRs	
		r	*p*-valuc	r	*p*-valuc
Disease severity	WHO-FC	-0,70	<0,01	-0,65	0,01
	PVRi	-0,67	<0,01	-0,75	<0,01
	mPAP	-0,64	<0,01	-0,67	<0,01
	mRAP	-0,46	0,02	-0,54	<0,01
	RV CI	0,43	0,02	0,61	0,01

P1986 - EXPLORING PARENTS' EXPERIENCES BRIDGING TO CARDIAC TRANSPLANT ON A BERLIN HEART

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Background: Devices such as the Berlin Heart, a mechanical circulatory support device, are increasingly being used to support children in cardiac failure whilst they wait for a suitable donor heart. Although the medical benefits of these devices have been established, studies have largely neglected the psychological impact of this experience on parents and their children. This study aimed to explore parents' experiences of supporting a child on a Berlin Heart as a bridge to cardiac transplant. Secondly, this study aimed to understand the impact these experiences had on parental quality of life, parental adjustment to their child's condition and family dynamics.
Methods: Semi-structured interviews were completed with eight parents of children who were supported on the Berlin Heart as a bridge to cardiac transplant. Interpretative Phenomenological Analysis was used to identify themes and connections across parents' accounts.
Results: Three superordinate themes were identified: 'Loss of Control on the Berlin Heart', 'Life in the Berlin Heart Bubble' and 'Transitioning Forward'. The findings highlight the complex process of decision-making for these parents and difficulties managing distress in a restricted and isolated environment. Loss of control was a defining feature of parents' experience. The importance of their support system in transitioning forward to a more positive and hopeful position with their child was also emphasised. There was a significant impact on parental quality of life and family dynamics throughout the experience.
Conclusion: The study provided a unique insight into the parental lived experience of having a child supported on a Berlin Heart through the collection of rich narrative data. There are a number of research and clinical implications for healthcare professionals including: how parents can be more adequately supported whilst their child is on a Berlin Heart, coping with uncertainty and the challenges of decision-making.

P2002 - PULMONARY HYPERTENSION AND ASSOCIATED FACTORS IN SICKLE CELL DISEASE

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Background: Sickle cell disease (SCD) affects ~100,000 people in the United States. Pulmonary hypertension (PHTN) is found in 6-11% of SCD adults and is associated with 40% mortality within 3 years. Less is understood about PHTN in the pediatric SCD population. Studies have estimated a prevalence of PHTN up to 30%, defined as tricuspid regurgitation velocity (TRV) ≥ 2.5 m/s on echocardiogram. The objective of this study is to describe PHTN in children with SCD.
Methods: Retrospective chart reviews were performed at an academic center caring for 450 children and adults with SCD. Subjects were eligible if they were 8 to 30 years during the period 7/1/2003 to 6/30/2015 and had their first screening echocardiogram before age 21. Patients with PHTN-predisposing comorbidities were excluded. Baseline hemoglobin (Hb), reticulocyte counts (RC), SCD-related complications, history of asthma, hydroxyurea (HU) use, and chronic blood transfusions (CBT) were assessed. Categorical and continuous data were analyzed using Fisher's exact test and 2-tailed t-test, respectively.
Results: Of 118 eligible subjects, 83 (70%) had a TRV measured (Table). Eleven of 83 (13%) had one or more TRV values ≥ 2.5 m/s. The average age at first elevated TRV was 15.9 years, and of these, nine (79%) had been prescribed HU at some point and 6 (55%) were prescribed HU at the time of elevated TRV. No statistically significant difference was found in TRV and genotype, Hb, HU use, SCD-related complications, asthma, or CBT. However, RC was significantly higher in the elevated TRV group.
Conclusion: Only 13% of patients with SCD had TRV ≥ 2.5 m/s, lower than published estimates. While other studies have found associations between TRV and genotype, Hb or SCD-related complications, these were not seen in this study. Prospective longitudinal studies beginning in childhood are needed to understand the development of PHTN among SCD patients.

Table. SCD-related factors associated with PHTN in previous studies

Factor	% of subjects with measured TR jet (N = 83)
HbSS	73
History of SCD-related conditions	
Acute chest syndrome	60
Splenic sequestration/splenectomy	22
Sepsis/bacteremia	17
History of Asthma	22
Hydroxyurea use	64
Chronic blood transfusion	8

P2003 - FRAGMENTED QRS COMPLEXES IN PEDIATRIC PATIENTS WITH IDIOPATHIC DILATED CARDIOMYOPATHY A SINGLE TERTIARY CENTER EXPERIENCE

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Background: Fragmented QRS complex (fQRS) is an easily evaluated noninvasive electrocardiographic parameter. Presence of fQRS has been associated with alternation of myocardial activation due to myocardial scar and myocardial fibrosis. Some literatures suggest that the parameter may affect prognosis and risk of sudden cardiac death, risk of implantable cardioverter-defibrillator therapy and response to cardiac resynchronization therapy. In this retrospective study we investigated the difference between presence and absence of fQRS.
Methods/Results: We retrospectively reviewed the records of pediatric patients <19 years of age who were diagnosed with idiopathic DCMP from 2003 to 2014 at Samsung Medical Center. Sixty-five patients were enrolled in this study according to the inclusion criteria: (1) pediatric idiopathic dilated cardiomyopathy patients aged <19 years, (2) performed the electrocardiography at diagnosis and recent follow-up. Median age at diagnosis was 14.5 months (range 1-237 months) and gender ratio was nearly 1.0 (male 32, female 33). In 25 patients (36/65, 55.4%), fQRSs were appeared at diagnosis. There were no statistically significant difference between fQRS positive and negative patients except the age at diagnosis (positive group: 81.49months; negative group: 41.28 months). In 41 patients (41/65, 63.0%), fQRSs were appeared at recent follow-up electrocardiography. Same as above, there were no statistically significant difference between positive and negative of fQRSs patients including mortality, cardiac death, cardiac contractility, and left ventricle dimensions. In our study, there were two cases of ventricular arrhythmia. One was a

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mortality case due to ventricular fibrillation, and the other case was frequent ventricular premature beats and non-sustained ventricular tachycardia. All cases of the ventricular arrhythmias, fQRSs were appeared.
Conclusion: Presence of fQRS is not always a strong independent predictor of clinical outcome in our experience. Despite of statistically non-significant findings, we will have to carefully observe the newly appeared ventricular arrhythmia in patients with fQRS.

P2005 - THE MID TERM RESULTS OF PEDIATRIC HEART TRANSPLANTATION

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Background: Heart transplantation (HTx) is a life-saving therapy for infants, children, adolescents and adults with end-stage heart failure or complex, inoperable congenital cardiac defects. Our team performed the first case of orthotopic HTx in July 1988 and first pediatric case (recipient's age under 18 years) in March 1996. By December 2016, a total of 467 cases have been performed with acceptable results. The aim of this study was to review retrospectively 21 pediatric patients who underwent orthotopic HTx. The mid-term survival results in pediatrics and adults were compared.
Materials and Methods: From July 1988 to December 2016, the total number of HTx cases was 467 in this program. Twenty-one of these 467 patients were pediatrics. Average age was 14.2 ± 3.5 years, ranging from 2 to 18 years, fourteen were male and seven female. The etiologies of end-stage heart disease are dilated cardiomyopathy in 15 patients, congenital heart disease in 4, and valvular cardiomyopathy in 2.
Results: Actuarial survival rates in pediatric cases at 1, 5, 10 years were 81.0%, 59.9%, and 28.1%, respectively. On the other hand, actuarial survival rates in adults at 1, 5, 10 years were 85.7%, 74.7%, and 59.5%, respectively.
Conclusions: Our results revealed that HTx is a feasible alternative for pediatric patients who suffered end-stage heart disease. With careful choice of the recipients and selection of appropriate immunosuppressive agents, durable mid-term result could be achieved. The actuarial survival rate in pediatrics after 5 years is relatively worse than that in adult cases, indicating the scarcity of pediatric donor hearts remains a serious problem and hemodynamic instability before HTx is relatively more common in pediatric cases. To improve the results, cooperation between the transplant team and the primary care pediatrician and well-organization of the pediatric donation resources are crucial.

P2020 - MYOCARDIAL PERFUSION ABNORMALITIES IN EISENMENGER SYNDROME

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Background: Compared to idiopathic pulmonary hypertension (IPAH), patients with Eisenmenger syndrome (ES) are known to have preserved right ventricular (RV) function for a longer period. While this difference is attributed largely to preservation of a fetal phenotype, the exact contribution of myocardial ischemia is not known.
Objective: We sought to examine the prevalence of myocardial ischemia in patients with Eisenmenger syndrome using stress nuclear studies.
Materials and Methods: In this prospective study, twenty consecutive patients with Eisenmenger syndrome were evaluated from 2014 to 2016. These patients underwent clinical evaluation, six minute walk test, echocardiography and stress myocardial scintigraphy using technetium 99 m sestamibi.
Results: Majority (60%) of patients were in were in NYHA functional class II. All patients had RV hypertrophy. Four (20%) patients had RV dysfunction while LV function was normal in all. Two (10%) patients showed perfusion defects in the anterior wall of right ventricle while one patient had perfusion defect in the interventricular septum. Four (20%) patients had perfusion defect in the LV apical area of whom it was reversible in two patients.
Conclusion: Myocardial perfusion defects both in RV and LV occur even in asymptomatic or mildly symptomatic patients with Eisenmenger syndrome. However, they are less frequent than reported in patients with IPAH, which may be partly responsible for the better prognosis observed in ES.

P2021 - APICAL RECOIL A SIMPLIFIED MEASURE OF UNTWIST MAY BE USED AS AN INDEX OF DIASTOLIC DYSFUNCTION IN CHILDREN WITH DILATED CARDIOMYOPATHY

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Backgrounds/Objectives: Untwisting of helical left ventricular (LV) myofibers during early diastole is defined as recoil. Recoil may be a precursor of LV apical "suction". Previously we identified apex as the "engine" of the LV. The purpose of this study was to simplify untwist by measuring recoil from a single view, the LV apex. We also evaluate the feasibility of utilizing apical recoil in children with dilated cardiomyopathy (DCM), to measure diastolic dysfunction.
Methods: Fifty-one healthy subjects (mean age 13.8 years age match + 2.9) were studied prospectively at rest and exercise. To increase lusitropy, subjects performed repeated straight leg raising from the hip to elevate their heart rates by > 40 beats per minute. Peak apical recoil rate and time to peak recoil were measured using 2DE speckle-tracking of LV apex. Application of this index in diseased states was tested in 18 DCM patients at rest (mean age 9.5 years, range 9 months to 18 years) with 30 sex and age matched control.
Results: In healthy subjects, peak apical recoil rate increased with exercise, -63.07 + 19.4°/s to -97.82 + 39.02°/s ($p < 0.01$) and time to peak recoil became shorter, 424.4 + 60.3 ms to

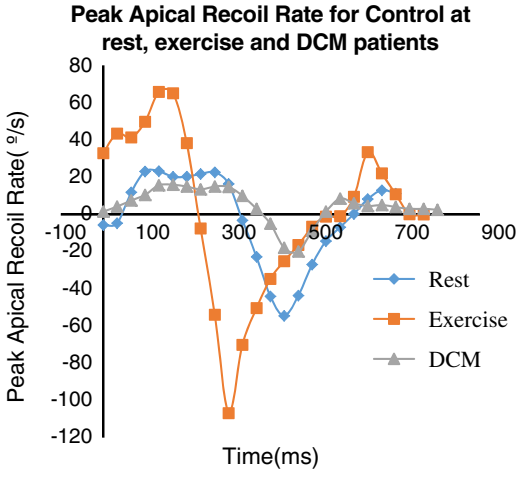

Figure.

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337.4 + 51.1 ms ($p < 0.0001$). Compared to controls, peak apical recoil rate was significantly lower in DCM cohort, -38.5 + 18.6 °/s vs -63.07 + 19.4 °/s ($p < 0.001$).
Conclusion: Apical recoil is new concept that may provide insight into LV untwisting and diastolic dysfunction. It was successful in detecting both increased and decreased lusitropic states. Commercial ultrasound machines could potentially automate this index for future clinical applications.

P2033 - ECMO THERAPY IN A CRITICAL PATIENT WITH TRANSPOSITION OF GREAT ARTERIES AND PERSISTENT PULMONARY HYPERTENSION

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Background/Hypothesis: Persistent pulmonary artery hypertension (PPAH) occurs in 1-3% of neonates with transposition of great arteries (TGA). This can dramatically increase mortality of TGA, due to persistent cyanosis and heart failure.
Material and Methods: We report a case where ECMO therapy was successfully employed to reduce pulmonary artery pressure and to improve oxygenation in a TGA-PPAH patient. A new born of 2,5 Kg with TGA presented severe desaturation and increasing levels of lactate, in spite of mechanical ventilation, prostaglandins, and inhaled NO. Rashkind atrioseptostomy didn't produce any changes in clinical situation. Echocardiogram showed a patent ductus arteriosus shunting right to left. Increasing dosage of adrenaline (1,5 mcg/Kg/min) was needed to maintain arterial blood pressure higher than 40 mmHg, while peripheral and cerebral saturation remained under 20%. To manage this critical situation, a venous-arterial ECMO (centrifugal pump) was implanted through cervical vessels (Arterial: 8 F, Venous: 10 F).
Results: After ECMO implantation, hemodynamic and respiratory function improved rapidly. Lactate levels normalized, and adrenaline requirements decreased. Echocardiogram showed a reduction of pulmonary pressure. We maintained a low dosage of prostaglandin in order to avoid the LV deconditioning. After 48 h of ECMO support, Arterial Switch Operation (ASO) was performed without complications. CPB weaning was made under low dosage of inotropes and no further mechanical assistance was needed in the postoperative period. Chest was left open prophilactically, and could be closed the 4th postoperative day. Patient was extubated the 12th day and discharged from ICU the 18th day. No complications were registered, except mild distal inferior limb ischemia. The patient was discharged home the 47th postoperative day.
Conclusion: ECMO therapy should be considered in TGA-PPAH critical patients, in order to improve the clinical and hemodynamic situation and to reduce pulmonary vascular resistances, so that ASO could be performed in optimal conditions.

P2107 - REPAIR OF ISOLATED ATRIAL SEPTAL DEFECT (ASD) IN INFANTS LESS THAN 12 MONTHS IMPROVES SYMPTOMS OF BRONCHOPULMONARY DYSPLASIA OR SHUNT RELATED PULMONARY HYPERTENSION

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Introduction: Infants with isolated ASDs are usually minimally symptomatic and repair is typically performed after infancy. Early repair may be considered in certain circumstances e.g. patients with reduced respiratory reserve or early signs of pulmonary hypertension (PH). Our aim was to review the characteristics and outcomes of a large cohort of infants who underwent infant ASD repair at our institution.
Methods: The study included 56 infants (28 female, 19 trisomy 21) with isolated ASD [age: 8 months (1.5-12), weight 6 kg (2.8-7.5), echo Qp/Qs:1.9 ± 0.1], who underwent surgical closure (20 fenestrated) between 2006-2016. Three groups of patients were identified: i) bronchopulmonary dysplasia of prematurity and PH (BPD-PH: n = 27) ii) Acutely unwell infants with PH but no chronic lung disease (non CLD-PH: n = 20, 36%) iii) Infants with refractory congestive heart failure (HF) without either PH or CLD (high-flow, n = 9, 16%). There were no major surgical complications. PH status was defined by echo (systolic PAP > 35 mmHg/ indirect markers).
Results: Postoperatively, PH infants (47/56) showed improvement in TAPSE z-score (BPD-PH:-1.74 ± 1.24 vs 1.04 ± 1; nonCLD-PH:-0.03 ± 2 vs 1.02 ± 0.1, $p < 0.001$) and right ventricular systolic/diastolic duration ratio (BPD-PH:1.37 ± 0.12 vs 1.1 ± 0.31; non CLD-PH:1.2 ±0.2 vs 0. 84 ± 0.1, $p < 0.05$). All ventilator (14,25%) or O2 dependent (31,55%) children could be weaned within two weeks after ASD closure. One year after repair, weight z-score increased in all patients and by +1 (0.5-1.5) in BPD-PH, +1.3 (0.7-1.7) in nonCLD-PH and +2 (1-2) in high-flow group. Over a median follow-up of 1.4 yrs (0.2-8.5), 3 patients died. Only 4 of 47 PH patients continued to have PH echo evidence at last follow-up. Only two of them remain on anti-PH medication.
Conclusion: This cohort suggests that ASD repair within the first year of life may improve the clinical status and growth.

P2127 - PHYSICAL ACTIVITY IN PEDIATRIC PULMONARY ARTERIAL HYPERTENSON MEASURED BY ACCELEROMETRY A CANDIDATE CLINICAL ENDPOINT

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Background: The development of evidence-based treatment guidelines for pediatric pulmonary arterial hypertension (PAH) is hampered by lack of pediatric clinical trials. Trial-design is hampered by lack of a feasible clinical endpoint in this population. We aimed to evaluate the use of accelerometry for measuring physical activity (PA) in pediatric PAH and to investigate its correlation with clinical disease severity markers.
Materials and methods: We included children from the Dutch National Network for Pediatric Pulmonary Hypertension. Controls were recruited from the outpatient cardiology clinic of the Beatrix Children's Hospital. Children were asked to wear the accelerometer for 7 days. Vector magnitude counts per minute (VM CPM) and time per day spent in different PA intensity levels were defined as accelerometer outcomes.

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Results: VM CPM was lower in children with PAH (n = 29) than in controls (n = 60) (647 vs. 921; $p < 0.001$) (Figure 1). Children with PAH spent less time in moderate and vigorous PA (13 vs. 29 minutes, 2 vs. 13 minutes per day; $p < 0.001$). Time spent in moderate and vigorous PA correlated inversely with WHO-functional class. Time spent in moderate PA correlated positively with 6-minute walk distance. In post hoc analyses, higher VM CPM and more time spent in moderate/vigorous combined and vigorous PA were associated with favorable outcome ($p \leq 0.044$).
Conclusion: PA is markedly decreased in children with PAH. Accelerometer output correlated with clinical disease severity markers and may predict outcome. We showed an exciting potential of PA as meaningful endpoint for clinical trials in pediatric PAH, although its clinical utility and prognostic value needs to be further validated.

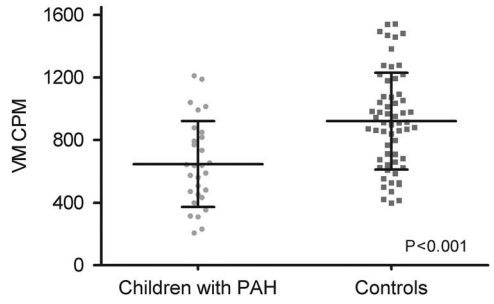

Figure.

P2128 - SYSTEMATIC LITERATURE REVIEW ON THE CLINICAL ECONOMIC HUMANISTIC AND SOCIETAL BURDEN OF HEART FAILURE IN CHILDREN AND ADOLESCENTS

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Background: Pediatric heart failure (HF) is associated with substantial morbidity and mortality. However, to date, there has been no systematic assessment of pediatric HF burden.
Methods: This narrative systematic review used Cochrane methodology to assess the evidence on the HF burden in children and adolescents (<18 years) over the past 10 years (2006–2016). Sources searched included Embase, Medline; selected congresses; and WHO, NHSEED, and HTA databases.
Results: 18 observational studies were included. Six studies reported mortality rates. In a UK/Ireland study, the 1-year mortality rate for patients admitted with a first HF episode was 18.3%, while a US study reported 71% mortality over 10 years for in-hospital HF following cardiac arrest. Based on three studies, 16.4%–82% of end-stage HF patients underwent heart transplantation. Four studies reported HF-related hospitalization rates of 18.2%–63.9% in infants (<1 year) and 18.7%–45.7% in children aged 1–12 years. Across six studies, mean length of hospital stay was 15 days overall, but was up to 26 days for infants. Five studies reported cost data. US studies reported mean hospital charges/year that were higher for infants (US$176,000) than for children aged 1—10 years (US$132,000) and total hospitalization costs for all US pediatric HF admissions in 2006 of US$753,149,082 (mean hospital cost per patient: US$49,354). In Germany, DRG-based hospital allowances per HF case were €3,498 in 1995 and €4,250 in 2009. In a Nigerian study, out-of-pocket expenses for HF treatment were 0.7%–122% of family income (mean US $17.6 ± 10.6/month); indirect costs (transportation/man hours) were 0.1%–57.1% of monthly income; and mean man-hour loss was 85.2 hours/year. No study reporting on patient quality of life fulfilled the selection criteria.
Conclusions: This review demonstrates the substantial burden of pediatric HF. However, the small number of studies, heterogeneity, and limited geographical coverage indicates the need for further research.

P2132 - REGIONAL SEPTAL HINGE POINT INJURY DRIVES ADVERSE BIVENTRICULAR INTERACTIONS IN PULMONARY HYPERTENSION

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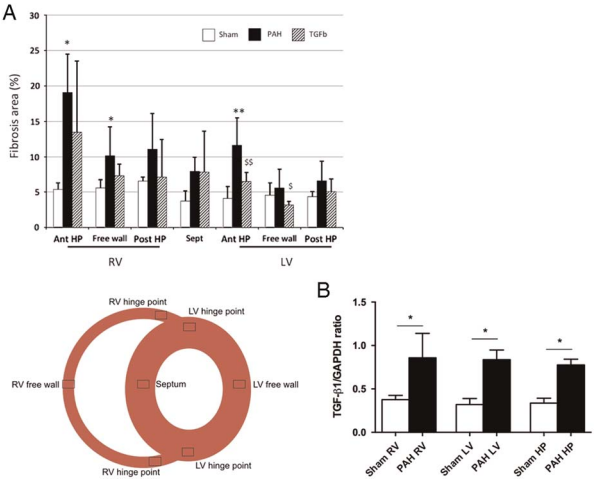
Background: Death and morbidity in pulmonary arterial hypertension (PAH) are often due to right ventricular (RV) failure and associated left ventricular (LV) dysfunction. We investigated regional myocardial remodeling and function as the basis for adverse ventricular-ventricular interactions in experimental chronic RV pressure overload.
Methods and Results: Two distinct animal models were studied: A) a rabbit model of increased RV pressure-load through progressive pulmonary artery banding B) A rat model of monocrotaline (MCT)-induced pulmonary arterial hypertension (PAH). Regional myocardial function was assessed by speckle-tracking strain echocardiography and ventricular pressures measured by catheterization before termination. Regional RV and LV myocardium was analyzed for collagen content, apoptosis and pro-fibrotic signaling gene and protein expression. Although the RV developed more fibrosis than the LV; in both models the LV was substantially affected (fig. A). In both ventricles, particularly the LV, fibrosis



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developed predominantly at the septal hinge-point regions in association with decreased regional and global circumferential strain, reduced global RV and LV function and up-regulation of regional transforming growth factor-β1 (TGFβ1) (Fig B) and apoptosis signaling. A group of PAH rats who received the TGFβ blocker SB431542 showed improved RV function and reduced regional hinge-point myocardial fibrosis.
Conclusion: RV pressure-loading and PAH lead to biventricular TGFβ1 signaling, fibrosis and apoptosis, predominantly at the septal hinge-point regions, in association with regional myocardial dysfunction. This suggests that altered geometry and wall stress lead to adverse RV-LV interactions through the septal hinge-points to induce LV fibrosis and dysfunction..

P2141 - STATUS OF PULMONARY ARTERY PRESSURES ON ECHOCARDIOGRAPHY IN HIGH RISK NEWBORNS

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Background: Persistent pulmonary hypertension of newborn (PPHN) is associated with various risk factors and there is paucity of data about the outcome of newborns who have one or more risk factors for PPHN and have some degrees of PAH not significant enough to cause hypoxemia and desaturation to be labelled as PPHN.
Objective: Prevalence of Pulmonary Arterial Hypertension in High Risk Newborns.
Material and Methods: This was a prospective observational study in which newborns (including both inborn and outborn deliveries) up to 1 month of age having risk factors for PPHN were included, a detailed history of mother and newborn was taken and after meticulous examination neonates having one or other risk factors for PPHN were selected for the study. Finally pulmonary hypertension was identified on echocardiography in neonates more than 48 hours of life with one or another risk factors. Second screening was done at 6 weeks of life independent of cases having PAH or not at the first screening.
Result: Total 410 neonates were screened and 10 cases were of congenital heart disease and hence excluded from the study. Out of remaining 400 cases 26 patients lost to follow up, 2 cases left against medical advice and 18 patients expired after first screening. Finally a total of 354 cases came for follow up at 6 weeks and repeat echocardiography was done in all cases. PAH was found in 54 cases on first screening, 12 neonates had reversal of shunt at the level of PDA or PFO and only 2 cases had persistence PAH on second screening at 6 weeks. Perinatal asphyxia (43.9%), respiratory distress (31%) and MSAF (30%) has shown a significant association with the development of PAH in these neonates.
Conclusion: Newborns with risk factors have some degree of PAH and should be screened early.

P2142 - INTERDISCIPLINARY APPROACH IN CHILDREN WITH PULMONARY HYPERTENSION (WHO GROUP 1)

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Background: Children with pulmonary hypertension (PH) require special expertise to diagnose and properly treat to improve long-term prognosis. Our Pediatric PH Unit is integrated by a team of pediatric pneumologists, cardiologists, neonatologists, intensivists, thoracic and cardiac surgeons and nurse practitioners experts in pediatric PH, with possibility to treat them with ECMO, atrial septostomy (AS) and lung transplantation (LT). We present our results of children diagnosed with pulmonary arterial hypertension (PAH): idiopathic or heritable PAH, associated with congenital heart disease (CHD) and veno-occlusive disease.
Main Methods: Retrospective, observational study. Data of our 27 children below 14 years with PAH controlled in our center from 2012 to 2014. The primary outcome was death, others outcomes were need for ECMO, AS or LT.
Results: We included 9 cases of idiopathic PAH, 16 of CHD-associated PAH, 1 veno-occlusive disease and 1 persistent PH. Mean age was 2.9 years(1 d-11.7 years; only 5 cases were older than 5 years); 15 (55.5%) females and 12 (44.5%) males. Eight children were diagnosed during this period, and the others were retrospective cases. AS was performed in 6(4 using diabolo fenestrated stent technique for mounting a stent; 1 by Amplatzer atrial septal-occluder with self-made fenestration and 1 was surgical). Time between diagnosis of PAH and AS was on average 20.4 months. ECMO was used in 3 (1 after LT) and LT was performed in 2. At follow-up 2 died (one at the moment of diagnosis and other after LT). The remaining 22 after a median follow-up time between 2 months and 14 years are stable on pulmonary vasodilator treatment (see figure).
Conclusions: To optimize the quality of life and the outcome of children with PAH, a specific interdisciplinary management is required. Strategies of physicians specialized on congenital heart diseases and PH should be warranted .

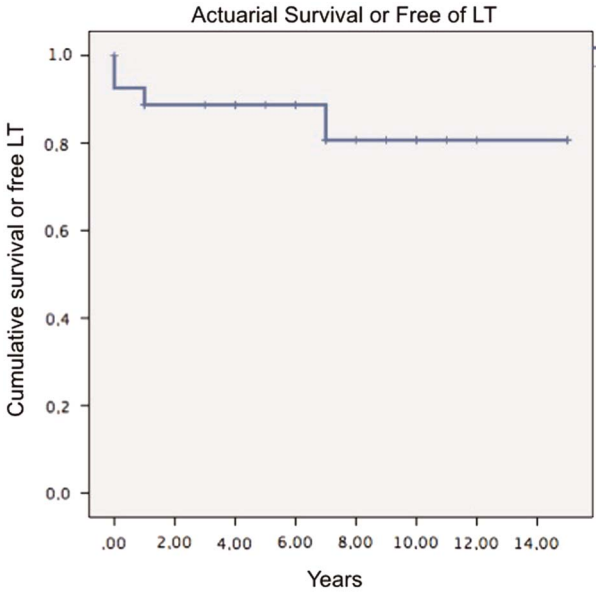

Figure 1.

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P2149 - PHARMACOKINETICS AND HEMODYNAMIC RESPONSES TO RECTAL TADALAFIL IN CHILDREN WITH PULMONARY HYPERTENSIVE VASCULAR DISEASE EARLY AFTER CARDIAC SURGERY

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Background: There is no report demonstrating the plasma concentrations of tadalafil in children with pulmonary hypertensive vascular disease (PHVD) after cardiac surgery. We assessed the pharmacokinetics and hemodynamic responses to rectal tadalafil for PHVD early after congenital cardiac surgery in pediatric patients.
Methods: Tadalafil is administered at a dose of 0.5 mg/kg every 12 hours rectally following admission to the intensive care unit. PHVD is defined as mean pulmonary pressure (mPAP) >25 mmHg for biventricular circulation and mPAP >15 mmHg or the mean transpulmonary pressure gradient (mTPG) >10 mmHg for single ventricle physiology. We retrospectively reviewed five patients treated with rectal tadalafil and measured tadalafil concentrations.
Results: Five patients with PHVD were included. Two patients were undertaken biventricular repair and 3 patients were undertaken bidirectional cavopulmonary shunt. The characteristics of patients were as below. Age:8.2 months (0.7-12.7), body weight: 5.3 kg(3.0-8.0), diagnosis: aortopulmonary window, complete atrioventricular septal defect (biventricular circulation)/pulmonary atresia with intact ventricular septum, tricuspid atresia, single ventricle (single ventricular physiology). Percentage of change in mPAP and mTPG significantly reduced eight hours after administration (-23.3 + /- 4.7%, $p = 0.03$) and further reduction was observed 12 hours after administration (-33.7 + /- 4.7%, $p = 0.005$). Systemic blood pressure did not change significantly during study period. Plasma concentrations of tadalafil increased gradually until second administration 12 hours after first administration. After second administration, the tadalafil concentrations increased further until third administration. The tadalafil concentrations were 48.0 + /- 9.2 ng/ml ($p = 0.019$) at 12 hours and 144.9 + /- 10.2 ng/ml ($p < 0.001$) at 24 hours after first administration. Moreover, there was negative correlation between tadalafil levels and percentage change of mPAP and TPG ($p = 0.01$, $r = -0.46$). No significant adverse events occurred.
Conclusions: Rectal tadalafil is a safe and effective treatment for children with PHVD early after cardiac surgeries.

P2218 - THE USE OF AN IMPLANTABLE CONTINUOUS FLOW VENTRICULAR ASSIST DEVICE IN A YOUNG SCHOOL AGE CHILD

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Background: The use of ventricular assist devices (VADs) as a bridge to transplant (BTT) in children with heart failure is expanding but still limited. We describe the use of an implanted continuous flow VAD as a BTT in a small pediatric patient who presented with dilated cardiomyopathy and cardiogenic shock.
Methods: An 8 year old 23 kilogram female with a body surface area (BSA) of 0.68 m^2 presented with diffuse abdominal pain, vomiting, and cough. Chest radiograph revealed massive cardiomegaly. Echocardiogram showed severe left ventricular dysfunction with an ejection fraction of 20%. She was admitted to a mixed pediatric cardiac intensive unit, initiated on inotropic support and evaluated for heart transplantation. Her precarious cardiorespiratory status deteriorated requiring intubation after which VAD support was decided. Four days after admission she underwent implantation of a HeartWare® ventricular assist device (HVAD). Device flows ranged between 2.2-2.4 revolutions per minute, providing an output of 2.9-3.6 liters/minute. The immediate postoperative course was complicated by a pericardial hematoma requiring mediastinal re-exploration and evacuation. Initial anticoagulation regimen included unfractionated heparin infusion. While being transitioned to an oral anticoagulation regimen with aspirin and warfarin her lactic dehydrogenase and plasma free hemoglobin levels were increasing. Due to concerns for hemolysis versus thrombus formation, the anticoagulation regimen was changed to bivalirudin and dipyridamole.
Results: She was extubated on postoperative day four after LVAD implantation. After 54 days on LVAD support she underwent orthotopic heart transplantation without complications and was discharged home on post-transplant day 11. She continues to do well, undergoing routine cardiac catheterization for biopsy with no evidence of rejection.
Conclusion: HVAD implantation was successful as a BTT in this young, thin patient without long-term complications or sequelae. Through the use of an expert multidisciplinary approach, detailed protocols and expedited decision making, these complex children can be successfully managed.

P2227 - ELECTRICAL CHANGES IN RESTING ECG IN CHILDREN WITH IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION

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University Children's Hospital Ulm, Division of Pediatric Cardiology, Ulm-Germany[1]; University of Giessen, Pediatric Heart Center, Giessen-Germany[2]

Introduction: Idiopathic pulmonary arterial hypertension (IPAH) is a rare condition in pediatric patients. Gold standard for diagnosis is invasive hemodynamic assessment. The role of electrocardiographic (ECG) changes in IPAH in children is still neglected. ECG ist widely available and easy applicable, even in small children. This study should systematically observe the role of ECG as a diagnostic and prognostic tool in pediatric IPAH.
Methods: Pediatric patients with suspected PAH who visited our tertiary referral center from 1999 to 2011 in order to obtain diagnostic cardiac catheterization were screened retrospectively. 42 children and adolescents were identified to have IPAH, a corresponding 12-lead-ECG was done during the same hospital stay. The follow-up after baseline examination was 59 ± 17 months. Kaplan-Meier analysis was used to assess predictive value of ECG changes regarding morbidity and mortality in these patients.
Results: In 12 patients (29%) severe cardiovascular events (defined as death, lung/heart-lung transplantation or Potts shunt) were observed after 20 ± 22 months. Patients with event showed higher baseline ratio of mean PA pressure to mean systemic arterial

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pressure (1.02 ± 0.26 vs. 0.84 ± 0.25, $p < 0.05$). Most patients (n = 37) showed a positive RV-Sokolow-Lyon index (RVSLI ≥1.05) at baseline ECG. Patients with event showed higher values (3.6 ± 1.2 mV vs. 2.6 ± 1.6 mV; $p < 0.05$). Relative risk for any event with an index more than double the common reference value (>2.1 mV) was 1.76 (95% Ci 1.21-3.20). Relative risk for death with RVSLI >2.1 mV was 2.01 (95% CI 1.61-4.80). RVSLI ≥2.1 mV was able to distinguish patients on risk for events with a sensitivity of 75% and a specificity of 56% (AUC 0.73, $p < 0.05$).

Conclusion: ECG with common reference values is not sufficient for staging and diagnosis of IPAH in children. IPAH-patients with elevated RVSLI >2.1 mV are at risk for cardiac events and should receive closer follow-up assessment, including repeated invasive evaluation allowing timely escalation of specific PAH-treatment.

P2237 - USING HEARTWARE DEVICE IN PAEDIATRIC PATIENTS WITH LEFT VENTRICULAR HEART FAILURE AS BRIDGE TO TRANSPLANTATION PROVIDES IMPROVED OUTCOMES AND BETTER QUALITY OF LIFE – UK BASED EXPERIENCE

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Background: The HeartWare Ventricular Assist Device (HVAD) is an implantable, centrifugal design, continuous-flow blood pump. Unlike EXCOR Berlin Heart, patients on HVAD are ambulatory and can be discharged home.

Method & Results: Retrospective review of 16 patients from 2 UK transplants centres [8 from each centre, mean age 11.3 (3-15) years, mean weight 39.3 (14-76) kg] underwent implantation of HVAD as left ventricular assist device, between 2014-2016, as bridge to transplant for end-stage heart failure. Median duration on HVAD was 187 (7-620) days. Table 1 shows the clinical background and outcomes following HVAD implantation. Of the 16 patients that received HVAD, there were no deaths. 15 patients have had successful transplantation and one remains alive on support. Most (13/16) children were discharged home, 2 were hospitalized long-term because of social reasons and 6/16 children returned to school. Complications following device implantation – 6 patients had driveline related infections requiring antibiotics. 3 patients had thrombo-embolic phenomenon causing reversible focal weakness but no patients have residual neurological deficit. 3 patients had sepsis with positive blood cultures requiring long-term antibiotics. One patient had thrombus in LV after VAD insertion and one had fungal infection with epistaxis and intracranial haemorrhage requiring neurosurgical evacuation. There were 2 pump blockades, one requiring emergency pump change and one transitioned onto cardiopulmonary bypass and urgent transplantation.

Conclusion: Our early experience shows that HVAD provides a relatively safe and efficient form of circulatory assistance for children awaiting heart transplant. Most of our patients on HVAD were able to go home, attend school and have community-based follow up, with overall good quality of life while awaiting transplantation. Further research is needed to prevent complications like infection, thrombo-embolism and haemorrhage in these patients..

Table 1. Demographics, clinical diagnosis and outcomes following HVAD implantation

Sr. No.	Age years	M /F	BW (kg)	Diagnosis	Total days on HVAD	Days of ICU stay	Outcome	Complications
1	15	M	36	DCM	87	13	OHT	Nil
2	10	F	40	Familial DCM	247	26	OHT	DLI
3	15	M		Chronic heart failure,	161		OHT	DLI, Sepsis
			54	PH		13		
4	14	F	36	DCM	546	5	OHT	DLI
5	12	F	37	DCM	255	33	OHT	Stroke, Sepsis (vegetations) Pump blockade
6	14	M	42	DCM [metabolic)	AT	35	-	Sepsis
7	15	F			68		OHT	Stroke; Bleeding; ICH;
			76	HCM		51		Pump blockade
8	11	M	33	DCM	224	7	OHT	Nil
9	3	M	14	Myocarditis	149	15	OHT	DLI
10	6	M	23	DCM	620	10	OHT	DLI; Stroke
11	7	M	18	D C M/ Myocar ditis	16	16	OHT	Nil
12	15	F	60	DCM secondary to Anthracycline	7	7	OHT	LV thrombus after VAD
13	12	F	52	DCM	50	12	OHT	Nil
14	4	F	14	DCM	201	7	OHT	Nil
15	15	M	50	OHT - graft rejection	275	29	OHT	DLI
16	13	M	44	HCM	187	6	OHT	Nil

OHT Orthotopic heart transplantation; DLI Driveline infection; ICH Intracranial haemorrhage; AT Awaiting transplant; PH Pulmonary hypertension; DCM Dilated cardiomyopathy; HCM Hypertrophic cardiomyopathy; LV Left ventricle;

P2245 - PEDIATRIC PALLIATIVE CARE IN ADVANCED HEART DISEASES

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Background/Hypothesis: Advance Heart Disease (AHD), continues to be a Life-Limiting condition and a leading cause of dead in pediatrics. Despite advances in the surgical and medical management of children suffering cardiac conditions little attention is paid to the end-of-life period. Uncertainties in prognosis and discrepancies in treatment goals between staff and family members have been described as barriers that interfere with optimal end-of-life care. The main objective was to analyze the model of care of patients attended by cardiologists and the Pediatric Palliative Care Team (PPCT).

Materials and Methods: Retrospective review of children with heart diseases attended by the PPCT in a tertiary care pediatric hospital from September 2009 to December 2016. Data is presented as Median (range).

Results: 18 patients with AHD (Structural Congenital Heart Disease (uni/biventricular), Cardiomyopathy, Arrhythmia and Pulmonary Hypertension) were attended. Median age: 6 years (8 days-18 years) at the time of palliative care consultation. Eight (45%) were females. Fifteen (85%) died [9 (60%) at home and 6 (40%) at the hospital]. The difference between survival times (1,48 month cardiac population and 5,8 months in the non-cardiac population) was not significant (p = 0,22)⋆. All children were

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surrounded by their parents at the time of death. Thirteen (72%) had comorbidities. PPCT activity was: 109 home visits, 150 inpatients visits and 467 phone calls. The primary care team was involved in 9 (50%) cases.
Conclusion: PPCT Facilitates The End-Of-Life Follow Up Of Patients Suffering AHD. In Our Series, A Multidimensional Approach Allowed Patients To Be At Home During The End-Of-Life Period Surrounded By Their Families. Future Studies Should Evaluate Patient And Family Perspectives Of Patients Dying From AHD In Order To Implement Strategies Directed To Ease The Healing Process Of A Children Death.

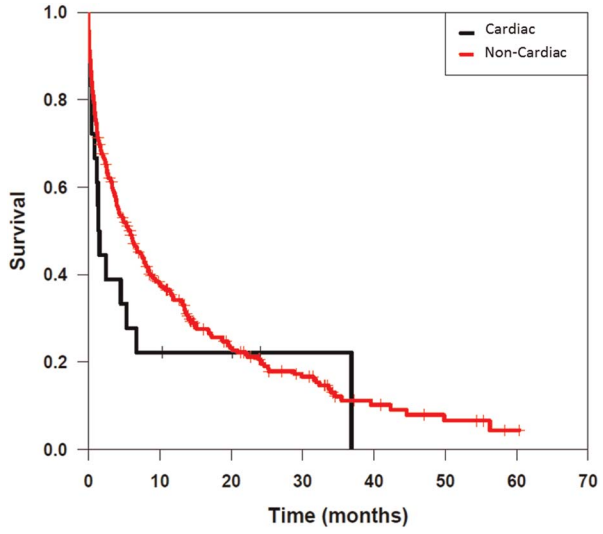

Figure.
Kaplan-Merier estimate of survival

P2270 - PEDIATRIC CARDIOMYOPATHY EXPERIENCE OF A UNIVERSITY HOSPITAL IN MARRAKESH MOROCCO

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Nicu, Mother and Child Hospital, Med Vi Hospital, Marrakesh Medical School, Cadi Ayyad University, Pediatrics, Marrakesh-Morocco[1]; Pediatrics A, Mother and Child Hospital, Med Vi Hospital, Marrakesh Medical School, Cadi Ayyad University, Pediatrics, Marrakesh-Morocco[2]

Background: Pediatric cardiomyopathy is a rare and severe heart condition that affects infants and children. Specifically, cardiomyopathy means disease of the heart muscle (myocardium).
Objective: To evaluate the frequency, diagnostic difficulties, the management and outcome of this disease. Materials and methods: A retrospective study of all cases of cardiomyopathy hospitalized in the pediatrics department at the university hospital Mohamed VI Marrakech, from January 2011 to October 2016.
Results: Cardiomyopathy was found in 32 of 7627 hospitalized children, which represents a prevalence of 0.4%. The mean age was 1.9 years (Extremes [1 month; 8 years]), with a male predominance (18 boys and 13 girls). All patients were symptomatic, 78% of them had heart failure. According to the ultrasound data: 26 dilated cardiomyopathy (DCM), 4 hypertrophic (HCM) and 2 case of restrictive cardiomyopathy (RCM). The etiological investigation confirmed a carnitine deficiency in one patient only, myocarditis was diagnosed in 41% of cases. Treatment was essentially symptomatic in all cases except in one case of DCM where treatment involved carnitine as well, one patient received a heart transplant. The Evolution was favorable under symptomatic treatment in 75% of cases with complications in 15% of cases, 9% of which resulted in death.
Conclusion: Cardiomyopathy is a rare, yet serious disorder of the heart that stands as a frequent cause of heart failure and the most common cause of heart transplantation in children, with substantial negative impacts on children and their families, as well as significant healthcare costs. The management of this disease is tough in Morocco because of financial problems, the difficulty for accurate diagnostic, the absence of some treatment in the country, and the lack of pediatric cardiologist.

P2300 - PREVALENCE OF PULMONARY ARTERIAL HYPERTENSION IN NEWBORNS WITH PERINATAL ASPHYXIA

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Background: Pulmonary arterial hypertension in adults is usually described as a pulmonary artery pressure of 25 mm of Hg or more. However neonates form a unique group in which there is a transition from systemic PA pressures to normal adult value and clear cut off value which is clinically significant is not well defined and situation gets more complicated when neonate have one or the other risk factor for pulmonary hypertension.
Objective: Prevalence of Pulmonary Arterial Hypertension in Newborns with Perinatal Asphyxia.
Material and Methods: This was a prospective observational study in which newborns up to 1 month of age having history and clinical features of perinatal asphyxia were included in the study. Both newborns delivered in our hospital and neonates delivered outside our hospital but admitted in our neonatal high dependency were enrolled in the study. Echocardiography was done after 48 hours of life and children with PAH were identified, second screening was done at 6 weeks of life independent of cases having PAH or not at the first screening.
Results: Total numbers of cases of perinatal asphyxia screened were 41 out of which 18(43.9%) cases had PAH. Out of 18 cases 11 neonates expired after first screening. A total of 7 cases came for follow up at 6 weeks. We also found that out of 18 neonates 8 had reversal of shunt at the level of PDA or PFO and hence were classified as cases of PPHN. Only 1 case had persistence PAH at 6 weeks screening.
Conclusion: Newborns with perinatal asphyxia had significant association with development of PAH which dramatically changes the prognosis as mortality was quite high in patients who developed PAH and even worse in those who developed reversal of shunt and systemic desaturation.

P2301 - ELIGIBLE CANDIDATE FOR LEFT VENTRICULAR ASSIST DEVICE AND OR HEART TRANSPLANTATION SUPPORTED WITH EXTRACORPOREAL MEMBRANE OXYGENATION IN CHILD AFTER CARDIAC SURGERY

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Background: Although extracorporeal membrane oxygenation (ECMO) has served as one of the effective care for children

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requiring mechanical support as a bridge-to-heart transplantation, initial indication of ECMO as a bridge-to-recovery might affect posttransplantation mortality in child of congenital heart disease after cardiac surgery. We aim to assess the morbidity and mortality in children requiring ECMO after cardiotomy and determine the eligible candidate for left ventricular assist device (LVAD) or heart transplantation.
Methods: Fifty-six infantile ECMO operations relating to congenital heart surgery were reviewed to analyze prognostic risks for the weaning of ECMO, the discharge of hospital and long-term survival were employed between January 2002 to December 2016.
Results: Age and weight were 13 ± 18months and 6.6 ± 3.3 kg, respectively. The mean follow-up was 5.6 ± 3.9year (0.4-13.6years). Twenty patients had single ventricle and 36 had biventricular physiology. The duration of ECMO was 8.1 ± 4.5 days. Forty-eight (85%) were successfully weaned off ECMO and 33 (59%) survived to hospital discharge. Eventually, 27 (48%) survived after hospital discharge. The weaning from ECMO was affected by peak serum lactate ≧ 60 mg/dl during ECMO (OR = 7.2, 95% CI: 1.5-1.8, p = 0.014) and the duration of ECMO ≧ 8days (5.0, 1.0-36.7, 0.046). Factors associate with failure of hospital discharge despite successful decannulation were as follows: weight ≧ 7 kg (0.24, 0.5-0.9, 0.024), ECMO duration ≧ 9days (3.6, 1.9-13.5, 0.041), the peak serum lactate ≧ 50 mg/dl of post ECMO (12.0, 1.5-5.2, 0.017). Ventricle physiology was not significant predictor. Survivor vs. non-survivor to the hospital discharge was 6.9 ± 4.1 vs. 9.8 ± 4.5 (day) in ECMO duration (p = 0.021). Post ECMO lactate ≧ 15 mg/dl was also affected in long-term survival.
Conclusions: Higher lactate levels more than 50 mg/dl and longer ECMO operation more than 8days are significant factors associated with mortality during ECMO and after successful decannulation. Patients with these conditions after cardiac surgery supported by ECMO are eligible candidates of LVAD and/or heart transplantation.

P2304 - PROFILE OF NEONATES WITH PULMONARY HYPERTENSION

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Background: Persistent Pulmonary Hypertension of Newborn (PPHN) is defined as a failure of normal fall in pulmonary vascular resistance (PVR) at or shortly after birth, leading to shunting of unoxygenated blood into the systemic circulation across foramen ovale or ductus arteriosus. Several risk factors are described which have association with PPHN in a newborn but there is little data on children with only PAH not severe enough to cause desaturation.
Objective: To study the outcome of neonates with risk factors having pulmonary hypertension on echocardiography.
Material and Methods: This was a prospective observational study in which newborns up to 1 month of age having risk factors for PPHN were included. Echocardiography was done after 48 hours of life and neonates with PAH were identified. Second screening was done at 6 weeks of life independent of cases having PAH or not at first screening. The spectrum and outcome these neonates was studied.
Result: Total numbers of neonates with PAH were 54 and out of these, 22(40.7%) cases were of Respiratory distress Syndrome (RDS), 18(33.9%) of perinatal asphyxia, 15(27.8%) of MSAF, 9 (16.7%) cases were of sepsis and twin delivery respectively. There were only 4(7.4%) cases of hypocalcemia, 3 cases were VLBW and 1 case of PIH. 12 cases had reversal of shunt and hence were classified as cases of PPHN. 13(24%) cases had expired of which 12 (22.2%) had perinatal asphyxia, 8(14.8%) had RDS, 3(5.5%) cases had MSAF and 4(7.4%) had sepsis. Neonates with multiple risk factors had the worst prognosis.
Conclusion: Various risk factors like RDS, perinatal asphyxia had significant association with development of PAH and mortality was significantly high in neonates with these risk factors after development of PAH.

P2318 - GROWTH DIFFERENTIATION FACTOR 15 AS A NOVEL BIOMARKER OF HEART FAILURE IN CONGENITAL HEART DISEASE WITH LEFT TO RIGHT SHUNT

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Background: Growth differentiation factor-15 (GDF-15) is a member of transforming growth factor–beta superfamily. Recent studies suggested that GDF-15 is an early diagnostic marker of heart failure in repaired congenital heart disease (CHD) and dysfunction of Fontan circuit. To our knowledge, there is no study about GDF-15 in unrepaired CHD.
Aim: To determine the efficacy of GDF-15 as a marker of HF with unrepaired CHD with left to right shunt.
Method: Plasma GDF-15 levels were measured in 45 patients (19 males) with left to right shunt CHD that received cardiac catheterization from March 2016 to December 2016. Median patients' age was 6 years old (range 0 - 71). Diagnosis was atrial septal defect in 28, patent ductus arteriosus in 11, ventricular septal defect in 4, coronary artery fistula in 1, and double outlet right ventricle in 1, respectively. From medical record, we determined the correlation between GDF-15 and clinical HF score (Ross and New York Heart Association classification), hemodynamic variables determined in catheterization, and NT-pro BNP.
Result: Patient's Ross/NYHA class was I in 40, II in 2, III in 3, and IV in none. Median concentration of GDF-15 was of 260 pg/mL (11-1024), NT-proBNP was 127 pg/mL (10-7922), and Qp/Qs was 2.1 (1.1-5.8). Mean SvO2 was 69.0 ± 5.8%, cardiac index was 3.0 ± 1.1 L/min/m2, and oxygen extraction ratio (OER) was 28 ± 5.6%. Because the distribution of NT-proBNP was not normal, NT-proBNP was log transformed. GDF-15 positively correlated with Ross/NYHA class (r = 0.54, p < 0.0001), Log NT-proBNP (r = 0.68, p < 0.0001), and OER (r = 0.48, p = 0.0008), but negatively correlated with SvO2 (r = -0.58, p < 0.0001). There was no correlation between GDF-15 with other hemodynamic variables including Qp/Qs.
Conclusion: Growth Differentiation Factor-15 can be an early diagnostic marker of heart failure in unrepaired congenital heart disease with left to right shunt in wide age range and different diagnosis.

P2328 - ECHOCARDIOGRAPHIC ASSESSMENT OF PAH AND CORRELATION OF ECHO PARMETERS WITH ITS SEVERITY AND SYMPTOMATOLOGY

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We present various echo parameters of the right heart from our institutional registry of patients with PH, evaluate right heart anatomy, hemodynamics and correlate with clinical features. Patient symptomatology, echo parameters of all patients were analysed for this study. 210 patients aged 27.415.7 years registered

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in our registry. Females dominated the registry. CHD associated PH constitutes (66.6%) and Idiopathic PH (30.4%). 72.2% were in NYHA II and 20.6% in NYHA III. Mean RVSP was 83.58. mean and diastolic PA pressures were 35.720 and 21.411.5 respectively. Estimated mean RA pressure was 12.5 + 8.9 mmHg. Increased RA area (>15.5 cm2) was seen in 76.4% patients and mean RA area was 20.81 cm2. Increased RV EDA >28 cm2 was seen in 38.7% and 36.4% had abnormal RV ESA >15 cm2.Mean FAC was 33 + 5.4% and 29% had FAC <32%. Abnormal RV strain <-18% was seen in 44% patients;mean strain was -17.73.74%. Abnormal PAT < 90 ms was seen in 39.2%; mean PAT value was 978ms. Pericardial effusion was seen in 13.8%. Study showed significant association of pericardial effusion with NYHA class III symptoms (p = 0.001), elevated RA pressure (p = 0.005, RA area >22 cm2 (p = 0.004), FAC <25% (P = 0.001), RV strain < -10% (p = 0.001), PAT <60 ms (p = 0.004). TAPSE <1.0 cm was associated with severe RV dysfunction (FAC 25 vs. 32%), right heart remodeling (RA area, 22 vs 15.5 cm2).Using logistic regression analysis,PE [p = 0.006,OR 4.03,95% CI(1.4-10.9)], RA area [p = 0.02, OR 3.1, 95% CI(1.1-8.0)] and PAT [P = 0.009, OR 0.34 95% CI(0.1-0.7)] were the 3 variables having significant correlation with symptoms. Our study identifies easily performed multiple echo parameters helpful in assessing the severity of PAH. With its close correlation to symptoms (NYHA class), echo parameters might be useful to assess outcomes, monitor the efficacy of therapy.

P2347 - OUTCOMES AND CHALLENGES IN 25 YEARS OF PEDIATRIC HEART TRANSPLANT IN A SINGLE INSTITUTION IN BRAZIL

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Background: Pediatric heart transplants (PHTX) continue to be the therapy of choice for children with end-stage heart failure. Since 1992, we have been experiencing challenges facing the interplay of limited donor supply and resources for mechanical circulatory support (MCS) and increase in growing-up congenital heart (GUCH) population. Recent reports suggest worse outcomes in congenital heart disease (CHD), specially in post-Fontan patients.
Hypothesis: Review our outcomes and risk factors in three distinct etiologies: cardiomyopathy (CMP), congenital heart disease (CHD) and Post-Fontan (s/p Fontan) patients. Casuistic and Methods: From 1992 to december 2016, 198 PHTX were performed in 190 patients:133 CMP (67,2%), 54 CHD (27,3%) and 11 post Fontan (5,6%) patients. The median age was 7,4 years (interquartile range 2,92-12,43 y), 49.7% male. Ten patients with GUCH diagnosis. Retrospective analysis was done using our medical record databank (Si3). Kaplan-Meier actuarial survival curve was obtained. Cox proportional-hazard regression method was used for univariate and multivariate risk analysis with 95% confidence interval using SPSS 23.0 (IBM,USA).
Results: Overall survival was 62,1% in 25 years. 30-day survival was 90,2%, 77,8% and 54,5% for patients with CMP, CHD and s/p Fontan respectively. One-year survival were 81,2%, 66,7% and 54,5% respectively and 10-year survival 71,4%, 55.6% and 54.5% respectively (log-rank p = 0,027). Re-transplants were done in 8 patients. The requirement for MCS after PHTX was greatest in patients with CHD (p = 0,001). Multivariate analysis revealed two significant risk factors for mortality: cardiopulmonary bypass time (hazard ratio[HR], 1.009; p = 0.027) and MCS as a bridge to PHTX (HR, 5.544; p = 0.003). CHD was a risk fator for early mortality (Odds Ratio 2.924, p = 0,006).
Conclusion: CHD and specially post-Fontan patients are higher risk groups in PHTX.

P2358 - THE PROGNOSTIC VALUE OF ECHOCARDIOGRAPHIC DIASTOLIC FUNCTION PARAMETERS IN PAEDIATRIC DILATED CARDIOMYOPATHY

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Background: Predicting the clinical course of paediatric dilated cardiomyopathy (DCM) remains challenging. Diastolic dysfunction is thought to be an important determinant for prognosis and survival in several paediatric heart diseases; however, its role has not been investigated in children with DCM.
Methods: We reviewed the first echocardiogram with diastolic function assessment for all children with dilated cardiomyopathy due to myocarditis, familial or idiopathic DCM from January 2006 to present. The association between diastolic function parameters and outcome was analyzed. Outcome was defined as recovery, stabilization and transplant or ventricular assist device for bridge-to-transplant. A univariable and multivariable analysis was performed using Cox regression to determine predictors of outcome.
Results: 44 echocardiograms of children with DCM were reviewed. The cause of DCM was familial for 8 patients, idiopathic for 26 and myocarditis for 10. Eight patients required cardiac transplant or mechanical assist devices (18%), 24 had persistently abnormal left ventricular function and/or dilatation (55%) and 12 patients recovered (27%). Median age was 1.7 years (range 0.17 to 16.08 years) and median duration of follow up was 3.0 years (range 0.17 to 8.6 years). Smaller values for lateral and septal E wave velocities on TDI and higher lateral E/E' ratios were associated with worse outcome (Table 1). Higher left ventricular end diastolic diameter z-scores (LVEDDz) and lower indices for left ventricular systolic function (fractional shortening (FS) and lateral S wave velocities on TDI) predicted for worse outcome. In a final multivariable model lower lateral S wave velocities were

Table.

	Univariable Analysis			Multivariable Analysis		
Variable	Hazard Ratio	95% CI	p-value	Hazard Ratio	95% CI	p-value
FS (%)	0.95	0.90-0.99	0.03			
LVEDDz	1.27	1.10-1.47	0.001	1.25	1.08-1.46	0.003
LVESD	1.05	1.01-1.09	0.01			
E/E'	1.05	1.0-1.10	0.02			
$E_{lateral}$	0.90	0.82-0.97	0.01			
E_{septum}	0.85	0.74-0.97	0.01			
$A_{lateral}$	0.90	0.74-1.10	0.31			
A_{septum}	0.98	0.90-1.06	0.6			
$S_{lateral}$	0.79	0.66-0.95	0.01	0.83	0.69-1.0	0.05
S_{septum}	0.81	0.65-1.02	0.08			
MV_A	0.79	0.16-3.80	0.77			
MV_E	0.99	0.98-1.0	0.06			
MV_{slope}	0.13	0.03-0.61	0.01			
E/A ratio	0.75	0.44-1.29	0.30			

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significantly associated with persistent or worsening disease (Hazard ratio, 0.83; p = 0.05) after adjusting for LVEDDz (Hazard Ratio, 1.25; p = 0.003).
Conclusions: Systolic function remains the most powerful predictor of outcome in paediatric DCM, but selected echocardiographic measurements for diastolic function can also help to predict outcome and warrant consideration in the assessment of these children..

P2360 - CARDIAC EXAMINATION IN CHILDREN WITH LARON SYNDROME TAKING MECASERMIN TREATMENT

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Introduction: Laron syndrome (primary growth hormone resistance or insensivity) is a rare genetic disease inherited autosomal recessive, although indistinguishable from growth hormone deficiency, it has high level growth hormone but IGF-1(insuline-like growth factor) and its related molecules can not syntesize.IGF-1 axis is important for cardiac development and cardiac protection. Mecasermin treatment is the only option for this patient. This study aims to research cardiac findings of children with Laron Syndrome taking mecasermin.
Materials and Method: The children with Laron Syndrome included in this study were diagnosed by applying growth hormone induction and IgGF-1 generation tests to children who have growth failure, at the pediatric endocrinology clinic. Their demografic data was obtained from their family and files. The children have taken mecasermin at 0,04-0,12 mcg/kg dose twice dubcutan injection per day, except for one child, who is going to take treatment after a six month follow-up. All of them were examined clinically, by electrocardiogram and echocardiogram.
Results: The study includes six children (4 male, 2 female). Three of them are siblings.Their ages were 5,76 + /-2,39 years. The treatment's duration was 8 to 53 months. Sistolic sufl was diagnosed in one child on the cardiac exam. They did not have arytmia. The echocardigrams did not show congenital cardiac anomaly, except patent foramen ovale in two patients. The echocardiogram of the child with sufl, showed pulmonary hipertension findings. These findings were proven angiographically. The patient's vasoreactivity test results were negative. Bosentan terapy was started. The reason of the ethiology of pulmonary hypertension is unknown.
Conclusion: In the literature we could not find pulmonary hipertension in childrin with Laron Syndrome, with or without mecasermin treatment. Cardiac findings and pulmonary pressures should be investigated on bigger series.

P2363 - AN OVERVIEW OF CURRENT PULMONARY HYPERTENSION TREATMENT IN SPAIN

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Backgound: Treatment options for pulmonary hypertension (PH) have evolved considerably in the past decade. However, the management of pediatric PH remains challenging as treatment decisions depend largely on experience of clinicians treating children. The aim of our study was to give a current view of PH treatment in our country.
Materials and Methods: A national, multicenter, retrospective, descriptive study was designed with the data collected in the Spanish Registry for Pediatric PH (REHIPED) about targeted therapies (TT).
Results: In April 2016, a total of 514 patients were registered in REHIPED (157 were dead, underwent lung transplantation or lost of follow up, and 357 were alive). Alive patients were classified into 2 groups: patients with a single ventricle physiology (Fontan group: mPAP <25 mmHg; n = 156; 44%) and patients with a biventricular circulation (PH group: mPAP >25 mmHg, PVRI >3 WU.m2; n = 201; 56%). Classification of Pulmonary Hypertension (WHO, Nice) in PH group was: 70% group I, 10%, group II, 17%, group III, 1% group IV and 2% and group V. TT were present in 75% of Fontan patients (117/156) and 71.6% of PH patients (144/201; 104 patients (73.8%) belonging to group I). Different treatments classified by groups are shown in the table. Monotherapy was present in 46% (n = 67) of PH patients treated, double therapy in 32% (n = 46, 72% Sildenafil + Bosentan; 78.7% group I) and triple therapy in 22% (n = 31; 39% Sildenafil + Bosentan + Treprostinil; 39% group I). In Fontan patients treated, 85% (n = 99) were in monotherapy, 14% in dual therapy (n = 17; 100% Sildenafil + Bosentan) and 1% in triple therapy (n = 1; Sildenafil + Bosentan + Iloprost).
Conclusions: In 2016 Spain, about 2/3 of patients received TT, most with Phosphodiesterase inhibitors. TT use is increasing in Fontan patients last years. Combined therapies were commonly used in PH patients.

Table.

Total patients	Phosphodiesterase inhibitors < n = 231;64J%)		EndotheEn recep tor antagonists < n = 114;31.9%)		Prostacyclins (n = 48; 11.2%)			CaJcruni Channel Blockers
Group Patients	Sildenafil	Tadalafil	Bosentan	Ambrisentan	Iloprost	Tieprostinil	Epoprostenol	
Fontan (n = 156)	103 (66%)	-	32 (20.5%)	-	1 (0.6%)	-	-	-
I (n = 141)	84 (59.5%)	8 (5.6%)	67 (45.5%)	4 (2.8%)	13 (9.2%)	17 (12%)	-	5 (3.5%)
II (n = 20)	13 (65%)	-	1(5%)	-	1(5%)	-	-	-
III (n = 34)	14 (41.1%)	3 (8.8%)	8 (23.5%)	-	5 (14.7%)	3 (8.8%)	-	-
IV (n = 2)	2 (100%)	-	1 (50%)	-	-	-	-	-
V (n = 4)	4 (100%)	-	1 (25%)	-	-	-	-	-

P2367 - USE OF PROSTANOID THERAPIES IN PEDIATRIC PULMONARY HYPERTENSION DATA FROM THE SPANISH REGISTRY

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Backgound: Prostacyclin therapy has formed the main stay of pulmonary hypertension (PH) therapy for many years and has improved functional status and survival for both adult and pediatric patients. The aim of our study was to describe the use of prostacyclins in children in our country.
Materials and Methods: A national, multicenter, retrospective, descriptive study was designed with the data collected in the Spanish Registry for Pediatric PH (REHIPED) about the use of prostanoids.
Results: In May 2016, a total of 492 patients were registered in REHIPED, 19% of them (n = 95) received /have received prostanoid therapy (PT). 3 cases were excluded for having univentricular circulation. Mean age at PH diagnosis was 3.3 ± 3.8 years (55% female). Mean time from diagnosis to start PT was 14.5 ± 25.5 months. Etiology Classification of Pulmonary Hypertension (WHO, Nice) included: 63% group I (41.3% idiopathic PH), 7.6% group II, 22.8% group III (66.6% bronchopulmonary dysplasia) and 6.5% group V (100% metabolopathies). Most patients (67%) before PT took 1 or 2 oral targeted theraphies (39% combined therapy, 28% monotheraphy) and 61% were in functional class III-IV. Distribution of PT by etiology groups is showed in the graphic. 7 patients received combined treatment with 2 prostanoids (85.7% Iloprost + Treprostinil). Most frequent adverse event with Iloprost was facial flush, Epoprostenol pulmonary edema and Treprostinil headache. Outcomes of PT after mean follow up of 3.9 ± 3.9 years were: 56.5% alive, 33.6% dead, 7.6% lung transplanted and 2.1% Potts shunt (n = 2; 1 alive; 1 dead). First cause of death was heart failure, followed by infection. Best survivals were found in group I and III, with no statistically significant differences.
Conclusions: In Spain, about 1/5 of PH patients have received prostanoids, most as a second/third line treatment. We have more experience in group I and with Iloprost and Treprostinil.

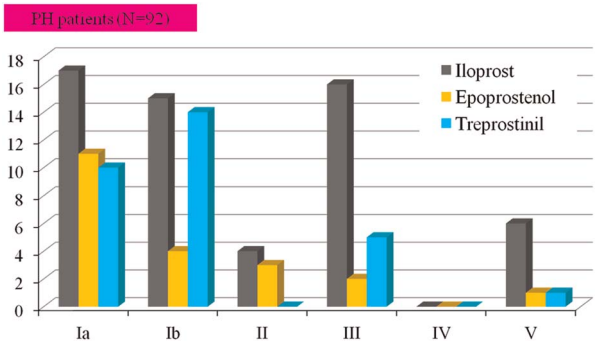

Figure.

P2395 - MECHANICAL CIRCULATORY SUPPORT STRATEGY OF BRIDGING TO PULSATILE FLOW IN PEDIATRIC PATIENTS IMPACT ON EARLY NEUROLOGIC EVENTS

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Background: Elective mechanical circulatory support (MCS) is used as triage as well as bridge to heart transplant (HTx). The incidence of neurologic (CNS) events, defined as having lasting motor or functional deficit, remains high, with clustering of events early post-implant when anticoagulation is not stabilized & post-operative bleeding predominates. Centrifugal MCS with Pedi or Centrimag (PM) support allows for less intense anticoagulation & a period of recovery immediately post-implant.
Methods: We compared our pts receiving MCS as triage or bridge to HTx: Group 1 (G1) pts received support from 1/10/2011 through 12/31/2014, N = 6; Group 2 (G2) pts received MCS 1/1/15 through 1/29/17, N = 6. G1 pts were placed on Berlin Heart Excor (BH) as chronic support until HTx (N = 6); G2 pts were placed on PM, if age < 15 yr, until adequately anticoagulated, then transitioned to BH (N = 5). Baseline demographics shown in Table 1.
Results: Incidence of CNS events in G1 was 67% (4/6) & in G2 was 33% (2/6). Time to CNS event in G1 was 7.75 d, range 1-11 d, & in G2 occurred at 4 & 9 d (6.5 d avg). Anticoagulation at CNS event in G1 & G2 was sub-therapeutic in all instances. Outcomes: In G1, all pts survived to HTx; in G2, 3/6 pts received HTx, 2/6 pts are waiting HTx on BH, 1 pt died after major CNS event while still on PM.
Conclusions: Elective MCS support with PM, as bridge to decision, HTx or BH conversion, shows promise resulting in fewer early CNS events in this small cohort of pts.

Table 1.

	Age (Avg, Med, R), yr	Diagnosis DCM: LVNC	CPR & ECMO prior to MCS	Atrial Communication at MCS implant	Normal CNS exam 24 hr prior to MCS	Listed 1st : MCS 1st	Duration PM Support (Avg, Med, R), d
G1	3.48, 0.38, 0.24-13.66	6:0	1	2, closed with ASD device	100%	5:1	X
G2	4.04, 0.39, 0.12-16.59	2:4	2	2, PFO < 1.5 mm	100%	2:4	13.8, 14, 11-16

AVG = average, Med = median, R = range; DCM = dilated cardiomyopathy; LVNC = left ventricular noncompaction; ASD atrial septal defect

Table 2.

	Age (Avg, Med, R), yr	Diagnosis DCM: LVNC	CPR & ECMO prior to MCS	Atrial Communication at MCS implant	Normal CNS exam 24 hr prior to MCS	Normal CNS exam prior to MCS	Listed 1st : MCS 1st	Duration PM Support (Avg, Med, R), d
G1	3.48, 0.38, 0.24-13.66	6:0	1	2, closed with ASD device	100%	6/6	5:1	X
G2	4.04, 0.39, 0.12-16.59	2:4	2	2, <1.5 mm	100%	6/6	2:4	13.8, 14, 11-16

AVG = average, Med = median, R = range; DCM = dilated cardiomyopathy; LVNC = left ventricular noncompaction; ASD atrial septal defect

P2404 - HEART RE TRANSPLANTATION IN CHILDREN AND ADULTS WITH CONGENITAL HEART DISEASE FOR GRAFT VASCULAR DISEASE 25 YEARS OF SINGLE CENTER EXPERIENCE

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Background: Heart transplantation is a therapeutic option for refractory heart failure. Graft vascular disease (GVD) is one of the limitations of long-term survival and may require new heart transplantation.
Patients and Methods: It was a retrospective observational study in cohort of children and adults with congenital heart disease submitted to heart transplantation between 1992 and 2017 at the Heart Institute (InCor) University of Sao Paulo Medical School. The incidence of re-transplantation and clinical outcome were analysed.
Results: Of the 190 patients who underwent heart transplantation, 6 required re-transplantation for GVD. The diagnosis of GVD was made by coronary angiography. The interval between the first and second transplants ranged from 9 to 19 years. The principal lesions were the right coronary artery (80%) and the anterior descending artery (60%). Two of the patients had renal failure as co-morbidity and required renal transplantation. One patient had renal transplant simultaneously and the other after cardiac transplantation. Two patients (33%) died in the immediate postoperative period after re-transplantation. The remaining patients (66%) were well and followed up at outpatient clinics.
Conclusion: The development of a coronary disease (GVD) is one of the main complications that limit the survival of cardiac transplant patients in the long term and re-transplantation is a successful therapeutic option.

P2411 - COMPARISON OF THE HEMODYNAMIC EFFECTS OF DOBUTAMINE AND NOREPINEPHRINE IN RIGHT VENTRICULAR PRESSURE OVERLOAD FAILURE

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Background: Right ventricular (RV) failure due to chronic pressure overload is an important determinant of outcome in congenital heart disease. Optimal inotropic therapy in the perioperative setting is challenging by the notion that not only contractility but also the interventricular interaction and arterio-ventricular relationship is manipulated. Several studies indicates that by increasing left ventricular (LV) afterload RV contractility can be improved, due to ventricular-ventricular interaction, hence improving cardiac index (CI).
Materials and Method: RV pressure overload was induced by pulmonary artery banding (PAB) for 16 weeks in 5 animals, and compared to 6 controls. Biventricular performance and hemodynamics were evaluated using biventricular conductance catheters, pulmonary catheter, and pressure transducers during infusion of norepinephrine (0,05–0,1–0,25-0,5 µg/kg/min) and dobutamine (0,5-1,0–2,5-5,0 µg/kg/min). We used a cross-over design and analyzed the results using repeated ANOVA analysis.
Results: After 16 weeks of RV pressure overload, Pmax was higher (45 vs 27 mmHg, $p = 0{,}001$) and Left ventricular volume (65 Vs 105 ml/m^2, $p < 0{,}001$), systolic pressure (78 Vs 98 mmHg) and cardiac index (CI) (4,0 Vs 5,4 L/m2/min, $p = 0{,}04$) were lower in PAB. During incremental inotrope infusions norepinephrine produced higher MAP ($p < 0{,}0001$) compared to dobutamine, increases in CI, HR and SV were equal between the groups. LV Arterial elastance (Ea) was significantly greater in norepinephrine treated PAB animals ($p > 0{,}001$), and the inotropic effect (PRSW) of norepinephrine in the LV was greater in PAB animals compared to control ($p < 0{,}05$), hence upholding the arterio-ventricular coupling and stroke volume.
Conclusion: In right ventricular pressure overload failure, with reduced LV volumes, CI and MAP, norepinephrine provided the same increase in CI but with higher MAP compared to dobutamine due to unchanged aterio-ventricular relationships in both ventricles.

P2417 - MECHANICAL CIRCULATORY SUPPORT IN PEDIATRIC HEART TRANSPLANTATION AND IN ADULTS WITH CONGENITAL HEART DISEASE SINGLE CENTER EXPERIENCE

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Background: Heart transplantation is the option therapy for patients with refractory heart failure. However, cardiogenic shock may occur before and after transplantation and the use of mechanical assistance circulatory support may be required. The objective of this study was the analysis of the use of mechanical cardiopulmonary support in pediatric patients and in adults with congenital heart disease before or after cardiac transplantation.
Patients and Methods: A retrospective clinical study of patients hospitalized with a diagnosis of pre or post transplant cardiogenic shock who used mechanical circulatory support (ECMO ™, Biopump ™ or Berlin Heart ™) from September 1999 to August 2016. Results: We included 44 patients submitted to mechanical circulatory support pre or after transplantation. The majority of patients were female (n = 26. 5%), aged 4 months to 27 years and weight of 5 to 48 kg. Short mechanical circulatory support (ECMO) was used in 28 (63.6%) patients, being in 16 (57.14%) of these as a bridge for transplantation. Biopump was installed in 17 (38.63%) and Berlin Heart in four (9%) patients. The shortest period in circulatory support care was 6 hours and the highest one was in one patient who used Berlin Heart for 334 days until transplantation. Heart transplantation was performed in 33 patients (75%) and 2 (4.5%) underwent re-transplantation. Renal insufficiency requiring dialysis, SIRS and stroke were the most frequent causes of death after transplantation, with the current survival of this group - 6 patients.
Conclusion: The installation of a mechanical circulatory support in patients with heart failure refractory to clinical treatment could be an option as a bridge for heart transplantation and helped to manage complications after the procedure.

P2419 - NOVEL NON INVASIVE ECHOCARDIOGRAPHIC MARKERS OF DISEASE PROGRESSION IN PEDIATRIC PULMONARY HYPERTENSION

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Background and Hypothesis: Pulmonary hypertension (PH) carries a poor prognosis due to eventual right ventricular (RV) failure. Although pulmonary artery systolic pressure (PASP) is used as a marker of PH severity, it is a poor prognostic marker of disease progression and for risk stratification. We sought to evaluate RV work from length-force relationship and pulmonary artery (PA) compliance and load as novel markers of PH disease progression in pediatric patients.
Method and Material: In a retrospective study, we analyzed echocardiographic indices in 20 children with confirmed PH (by catheterization). We measured tricuspid annular plane systolic excursion (TAPSE) for length shortening to assess RV systolic function, tricuspid regurgitation (TR) jet velocity to estimate PASP, and PA acceleration time (PAAT) to evaluate PA compliance and resistance. TAPSExPASP and TAPSExPAAT were calculated to develop length-force relationship as indices of RV work. The children were divided into two groups based on who reached a composite endpoint (intravenous pulmonary vasodilator, Potts shunt, lung transplant, or death) (group 1, n = 8) and those who did not (group 2, n = 12). Comparisons of the echocardiographic indices were made to an age/weight- matched controls (n = 11), and ANOVA was used to compare the indices amongst the three groups.
Result: TAPSE, PAAT, TAPSExPAAT were all significantly reduced in PH patients who reached the composite endpoints compared to PH patients who did not reach the endpoint and controls ($p < 0.01$ for all). Although PASP and TAPSExPASP were significantly reduced in PH patients when compared to the controls ($p < 0.01$ for both), they were similar between two PH groups ($p = 0.43$ and $p = 0.99$) (Table 1).
Conclusions: TAPSExPAAT as indices of RV work and PAAT as an index of PA load and compliance offer quantitative markers of disease progression in pediatric patients with PH that may be useful for risk stratification for intervention.

Table 1. Comparison of Echocardiographic Measures

		Pulmonary Hypertension		
	Pediatric Controls (n = ll)	**Do not reach endpoints* (n = 12)**	**Reach endpoints* (n = 8)**	**P values**
RV function TAPSE (cm)	2.0 (0.1)	1.6 (0.3)§	1.3(0.1)**†	p < 0.001
Pulmonary Hemodynamics				
PASP (mmHe, derived byTR jet velocity)	20‡	62 (27)§	76 (24)	p = 0.43
PAAT (msec)	127(16)	90(15)§	65(13)** †	p < 0.001
RV Work (length x force)				
TAPSE x PASP (cm x mmHg)	41 (2)	96 (46)§	96(25)†	p = 0.003
TAPSE x PAAT (cm x msec)	262(43)	152(40)§	82(16)** †	p < 0.001

Values are presented as mean (standard delation). One way ANOVA was used to compare the measures amongst the 3 groups
R.V. right ventricle; TAPSE. tricuspid annular plane systolic excursion (cm)
‡PASP. pulmonary artery systolic pressure. PASP was assumed to be normal for controls (~20mmHg) TR; tricuspid regurgitation (meters/sec); PAAT. pulmonary artery acceleration time (msec)

★Composite endpoints. defined as me need for intravenous medication, hang transplantation. Potts shunt, or death
** $p < 0.05$. PH patients who reach composite endpoint compared with PH patients who do not reach endpoints;

§$p < 0.01$. PH patients who do not reach composite endpoint compared with pediatric controls;
†$p < 0.05$ PH patients who reach composite endpoints compared with pediatric controls

P2428 - HEART TRANSPLANTATION IN CHILDREN AND ADULTS WITH CONGENITAL HEART DISEASE CHARACTERISTICS OF EFFECTIVE DONORS – SINGLE CENTER EXPERIENCE

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Introduction: Cardiac transplantation is a safe and effective alternative in the treatment of refractory heart failure. In order to ensure the integrity of the organ obtained and reduce risks to the recipient, some criteria must be respected.
Objective: To evaluate the characteristics of donors accepted for children for heart transplantation as well as the refusal of potential donors. Methods: A retrospective descriptive study was carried out in which the records of potential donors for children and adults with congenital heart disease were evaluated from October 2014 to August 2016 for anthropometric, clinical and laboratory findings.
Results: A total of 225 donors, aged between 6 months and 59 years, of which 118 (52.44%) were male. The main causes of donor brain death were: hemorrhagic stroke in 86 (38%) cases, trauma in 76 (34%), encephalopathy pos- cardiac arrest in 15 (7%), bacterial meningitis in 8 (4%) and Firearm accident in 8 (4%). Among the main criteria for refusal of the organs were: incompatibility in 76 (34%) cases, echocardiogram and/or electrocardiogram abnormalities in 71 (32%) cases, sepsis in 40 (18%), systemic arterial hypertension in 36 (22%), the occurrence of cardiac arrest in 23 (10%),hemodynamic instability in 22 (10%), difficulties related to logistics (distance between cities, availability of aircraft and climatic conditions for landing and takeoff) on 16 (7%) occasions, in 8 cases (4%) due to virtual positive cross-match and other issues (11%). Twenty-nine transplants were performed in this period, making a 13% utilization rate of the organs offered.
Conclusion: The majority of donors were male, died due to central nervous system diseases, of vascular or traumatic causes and the main criteria of refusal of the organs were anatomical disproportion, cardiac function alterations and sepsis.

P2434 - AUTOMATIC IMPLANTABLE CARDIOVERTER DEFIBRILLATOR IN CHILDREN WITH DILATED CARDIOMYOPATHY

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Background: Sudden cardiac death (SCD) is a known risk in patients with dilated cardiomyopathy (DCM). Predictors of SCD in children with DCM are not well defined. In children with DCM we looked for predictors of fatal arrhythmias and efficacy of automatic implantable cardioverter-defibrillator (AICD).
Methods: From 1999 to 2011,out of 300 children with heart failure, we identified 100 children with DCM. Medical records were reviewed for demographic information, New York Heart Association (NYHA) or Ross functional class, left ventricular ejection fraction (LVEF), left ventricular diastolic diameter dimension indexed by BSA (LVDDz), NTproBNP levels and outcome. The patients were divided into 2 groups: Group 1, with no AICD implantation; Group 2, with AICD implantation.
Results: Mean age of presentation was 7.6 ± 6.6 years, mean follow-up was 6 ± 5.3 years. AICD was placed in 23 patients (23%) because of ventricular tachycardia. The median time from presentation to AICD placement was 667 days. There was no difference in sex, ethnicity or race between the two groups. The age of presentation was significantly higher in Group 2 (Table-1). There was no significant difference in LVEF, NTproBNP,

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LVEDDz and NYHA/Ross class at presentation. At the last follow up (endpoint), NYHA/Ross class, NTproBNP and LVEF were significantly worse in Group 2 compared to Group 1 (Figure-1). Significantly higher number of patients in Group 2 required heart transplant. In the patients with AICD, there were 56 appropriate shocks in 10 patients and 9 inappropriate shocks in 7 patients.
Conclusions: Based on the information analyzed, we were not able to distinguish patients at risk of arrhythmia at time of initial presentation. Patients requiring AICD placement were older and showed worsening LVEF, NYHA/Ross class and higher NTproBNP, reflective of more severe disease. AICD can effectively rescue patients from VT and avoid sudden cardiac death; the number of inappropriate shock was small.

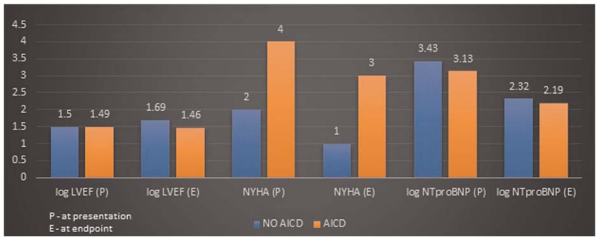

Figure.
Heart Failure Parameters in Children with and without AICD

Table 1. Comparison of Demographic and Clinical Information

Patient characteristics		Group 1 (No AICD) N = 77	Group 2(with AICD) N = 23	p-value
Age[1]	at presentation	3 (19)	14 (16)	0.001
Sex[2]	Mae	38 (49)	13 (56,5)	0.621
	Female	39 (51)	10 (43.5)	
Ethnicity[2]	Hispanic	32 (41.6)	9 (39.1)	0.833
	Non -Hispanic	44 (57.1)	14 (60.9)	
Race[2]	Caucasians	42 (54.5)	12 (52.2)	0.483
	African-Americans	31 (40.2)	11 (47.8)	
Outcome[2]	Death	8 (10.3)	3 (13)	0.721
	Transp ant	9 (11.7)	14 (60.9)	0.001
LVEF	at presentation[3]	3133 (±13.08)	3064 (± 11.42)	0.830
	at end point[1]	49 (61)	29 (50)	0.001
LVEDDz	at presentation[3]	4.57 (±3.04)	3.53 (± 2.00)	0.174
	at end point[1]	2.99 (17.38)	3.42 (9.74)	0.309
NYHA[1]	at presentation	2 (3)	4 (3)	0.138
	at endpoint	1 (3)	3 (3)	<0.001
NTproBNP[1]	at presentation	2713 (56571)	1341 (3370.1)	<0.001
	at endpoint	206 (39113)	1550. (8951)	0.004

1: Median (Range/, 2: Number (Percent), 3:Meon (SD)

P2441 - MANAGEMENT OF PULMONARY HYPERTENSION ASSOCIATED WITH A LARGE PATENT DUCTUS ARTERIOSUS IN EXTREMELY PRETERM INFANTS WITH BRONCHOPULMONARY DYSPLASIA

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Background: Children born premature often have a patent ductus arteriosus (PDA), which in association with broncho-pulmonary dysplasia (BPD) in these patients leads to early onset pulmonary hypertension (PHT). The objective of this study was to evaluate the hemodynamics and determine the effects of pulmonary vasodilation and PDA closure in these patients.
Methods: Premature infants that had severe PHT, defined as a pulmonary artery systolic pressure (Pap) > 50% of the systolic blood pressure (SBP), with severe BPD (ventilator and oxygen dependency) and a large PDA were evaluated over a 4-year-period. Those with another level of left-right shunting were excluded. All patients underwent cardiac catheterization. Hemodynamic assessment at baseline, followed by pulmonary vasodilatory therapy with and without test occlusion of the PDA was performed.
Results: Twenty patients born prematurely (26.2 ± 3.5 weeks' gestation, birth weight = 739 ± 324 grams), underwent cardiac catheterization at 102.6 ± 50 days of age, weighing 2.1 ± 0.68 Kg. At baseline ventilator support, the Qp:Qs was 1.5 ± 0.5, Pap was 74 ± 14% of SBP, and PVR was 3.94 ± 2 Wu•m2. With 100% oxygen and inhaled nitric oxide, the Qp:Qs increased to 2.6 ± 1, Pap decreased to 54 ± 10% of SBP, and the PVR decreased to 1.9 ±1.2 Wu•m2. At this stage when the PDA was test occluded, the shunt was eliminated with further decrease in the Pap and PVR (in all but one patient). Based on these findings, the PDA was closed in nineteen of the twenty patients.
Conclusions: Pulmonary vasodilator therapy for PHT increases the PDA shunt in the premature infants which could worsen BPD. Therefore, closure of the PDA could be beneficial in the effective management of PHT and BPD in these patients. Transcatheter PDA closure can be accomplished during the time of the hemodynamic catheterization.

P2468 - HEART TRANSPLANTATION IN CHILDREN AND ADULTS WITH CONGENITAL HEART DISEASE POST TRANSPLANT LYMPHOPROLIFERATIVE DISORDER – 25 YEARS OF SINGLE CENTER EXPERIENCE

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Background: Heart Transplantation has been the option for treatment of patients with refractory heart failure or inoperable congenital heart disease. However after the procedure, one of the main complications is post-transplant lymphoproliferative disorder (PTLD). The aim of this study is to report the incidence of PTLD after heart transplantation using double immunosuppressive maintenance therapy.
Patients and Methods: It is a retrospective observational study from a cohort of 190 patients who were submitted to heart transplantation at the Heart Institute (InCor) University of Sao Paulo Medical School from 1992 to 2017. The immunosuppressive therapy was calcineurin inhibitor and citostatic drugs. The diagnosis of PTLD was made by pathology findings and imaging methods.
Results: 21 of 190 (11%) patients developed PTLD. 7 (33%) were male. The sites of the disease were: 9 (42.8%) cervical lymph nodes, gastrointestinal 7 (33%) and 4 in the lungs. The treatment consisted of reduction of immunosuppressive drugs, the use of rituximab and sirolimus. The survival was 71.4%. The causes of mortality were 5 (83.3%) infection and 1 sudden death.
Conclusions: PTLD is a challenging complication after transplantation. The management of immunosuppressive drugs improved the survival.

P2481 - ANEURYSMATIC FISTULAS IN PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM IN CHILDREN CANDIDATES FOR HEART TRANSPLANTATION

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Introduction: The presence of anomalous coronary pathways has been seen in pathological studies of children for more than 60 years. There is a broad spectrum of coronary lesions in patients with pulmonary atresia with intact ventricular septum, the most common being the presence of ventriculo- coronary fistulas observed in 45% of the cases. The objective of this study is to report two patients diagnosed with pulmonary atresia with intact ventricular septum with coronary sinusoides in the postoperative period of Glenn's surgery and Blalock-Tausing surgery with aneurysmal dilatation of the right and left coronary arteries and candidates for heart transplantation.
Case 1: Female patient, 14 days old, performed Blalock-Taussig due to pulmonary atresia with intact septum and right coronary fistulas dependent on the right ventricle listed for cardiac transplantation. She developed heart failure and arrhymthymia (bigeminism). Cardiac transplantation was performed in 1993 and aneurysms of right ventricular coronary fistulae were detected during heart transplantation procedure. Patient is clinically well after 23 years and 10 months after heart transplantation.
Case 2: A 17-year-old female patient with a diagnosis of intact septum pulmonary atresia and late postoperative Glenn surgery referred for heart transplantation due to significant ventricular dysfunction and listed for heart transplantation. Coronary artery angiography shows the presence of a tortuous pathway of the right and left coronary arteries, with an aneurysmal dilatation of the left artery measuring 26 mm, visualized in the coronary sinus, left and right coronary arteries, and directly communicating with the right ventricle cavity.
Conclusion: Coronary aortic aneurysmatic fistulas can be severe and may evolve with the need for heart transplantation.

P2488 - TEMPORARY RIGHT VENTRICULAR ASSISTIVE DEVICE (RVAD) POST LEFT VENTRICULAR ASSISTIVE DEVICE (LVAD) IMPLANTATION

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Introduction: Mechanical circulatory support as either bridge to transplant or destination therapy remains a viable option for heart failure in pediatric patients. While outcomes shows improving survival rates, the management of right ventricular failure (RVF) after Left Ventricular Assistive Device (LVAD) implantation remains a challenge (Mansfield et al, 2015).
Background: Our patient was a previously healthy 27 kg, six year old male, newly diagnosed dilated cardiomyopathy with an ejection fraction of 25%, positive for mutations in MYH6 and LMNA and with no active inflammation on MRI. Heartware LVAD implantation was performed following ECMO deployment secondary to decompensation. Patient subsequently developed RV failure that necessitated rotaflow RVAD support.
Discussion and Implication for Clinical Practice: Pulmonary vasodilators, afterload reducers and the use of inotropes do not guarantee success in preventing the progression of RVF. The use of temporary RVAD may be considered to support medical management. Various options such as Berlin Heart, Total Artificial Heart, Heartware RVAD were considered but decided to be unfavorable on this case. Among the challenges met on a patient with temporary RVAD and Heartware LVAD are neurologic complications specifically Horner's syndrome, a nerve injury due to cannula placement, arrhythmia and frequent suction events on LVAD. Patient's mobility limitations required a complex multidisciplinary planning. Also, the rotaflow is prone to fibrin and clot formations making anticoagulation management difficult. After 45 days, patient was able to completely come off RVAD and two days later was successfully transplanted.
Nursing Implications: The patient's mobility restrictions, skin issues, suction events, unfamiliarity on the use of the rotaflow device and educational needs in managing both devices posed as some of the nursing challenges. The success of this case points to the strong collaboration among the multidisciplinary team, the development of educational tools and strategies, the enthusiasm and the strong conscientious work of all involved.

P2489 - THE EPIDEMIOLOGY OF CHILDHOOD DILATED CARDIOMYOPATHY IN SINGAPORE

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Introduction: The epidemiology of dilated cardiomyopathy in children is not well established. Little is known about the causes, outcomes and overall survival of childhood dilated cardiomyopathy in Singapore. Hence, we undertook a retrospective study to document the epidemiology of dilated cardiomyopathy in children who presented to a tertiary children's hospital in Singapore.
Methods: This was a single-centre, retrospective review of all cases of dilated cardiomyopathy in children who presented between 2005 and 2015 to the cardiology service at KK Women's and Children's Hospital, Singapore. All diagnoses of dilated cardiomyopathy were made using echocardiographic criteria.
Results: A total of 43 dilated cardiomyopathy patients were analyzed. There was a slight male predominance (n = 24, 56%) among the children who were diagnosed. The commonest race included Chinese (n = 23, 54%), followed by Malay (n = 7, 16%) and Indian (n = 3, 7%). The mean age at diagnosis was 5.4 years. Most children presented before the age of 6 years (n = 29, 67%), with 37% being diagnosed before 1 year (n = 16, 37%). The majority of children had idiopathic dilated cardiomyopathy (n = 18, 42%). Out of the known causes, most were secondary to neuromuscular disorders (n = 8, 19%) and myocarditis (n = 7, 16%). The mean follow-up time was 3.8 years. The mortality rate in this cohort was 30% (13/43). In multivariable analysis, age at diagnosis <1 year, and lower baseline left ventricular fractional shortening were associated with increased risk of death.
Conclusion: Without the availability of pediatric heart transplantation, death occurred in 19% of patients with childhood dilated cardiomyopathy within 1 year of diagnosis. Risk factors for death include age at diagnosis and severity of left ventricular dysfunction. This data is pertinent as Singapore moves to develop a pediatric heart transplant program.

P2504 - RETROSPECTIVE STUDY OF OPTIONAL TREATMENT STRATEGY USING PULMONARY VASOREACTIVITY IN BORDERLINE EIGENMENGER SYNDROME USING PULMONARY VASOREACTIVITY

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Background: This study aimed to evaluate the feasibility, clinical application of pulmonary vasoreactivity testing with inhaled iloprost and to determine the treatment strategy in patients with pulmonary arterial hypertension (PAH) associated with congenital heart diseases (CHD).
Method: This retrospective study was done in single center for 139 cases with CHD diagnosed as PAH through the right heart catheterization from Jan. 2011 to Jul.2016. Inhaled iloprost was delivered during cardiac catheterization by ultrasonic nebulizer via a flow-inflating bag with 0.02 or 0.04 mg. The hemodynamic results and post-catheterization treatment results were reviewed.
Results: The median age of the patients was 32.48 years. The average numbers of systolic pulmonary artery pressure (sPAP) and mean pulmonary artery pressure (mPAP) were 67.39 ± 35.77 mmHg and 44.39 ± 26.30 mmHg. The median pulmonary vascular resistance index was 7.42 Wood units. Among those patients, 58 patients had underwent the pulmonary vasoreactivity test with inhaled iloprost plus oxygen during catheterization (testing group). The average numbers of sPAP and mPAP were 88.32 ± 22.77 mmHg and 57.34 ± 18.37 mmHg in testing group. The median pulmonary vascular resistance index (PVRi) was 10.8 Wood units. 32 (55%) patients were regarded as responder group. The mean PVRi was also significantly decreased in responder group (9.6 to 5.6 vs 12.3 to 11.0 Wood units). After pulmonary vasoreactivity testing, 27 patients had undertaken complete/partial corrective procedure, 4 patients had undertaken one more procedure/operation followed initial procedure after catheterization, and in only 1 patient, palliative coronary stenting was done. The others were treated with single or combined pulmonary vasodilators without any procedure/operation.
Conclusion: We suggest that even in patients with PAH associated with CHD, pulmonary vasoreactivity test in RHC is essential to determine the treatment strategy of closure or not for borderline Eigenmenger syndrome.

P2506 - CAUSES OF HEMOPTYSIS IN EISENMENGER SYNDROME – A CT ANGIOGRAPHY STUDY

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Background: Hemoptysis is a common cause of morbidity in Eisenmenger syndrome, but the causes of hemoptysis are not well defined. We analyzed the clinical predictors and causes of hemoptysis in a cohort of patients with Eisenmenger syndrome using computerized tomographic pulmonary angiography (CTPA).
Materials and Methods: Of the 95 patients of Eisenmenger syndrome studied (mean age 23.7 ± 7.7 years; 57 male), 38 patients (40%) had presented with hemoptysis, and all of them underwent a CTPA within two weeks of index bleed.
Results: Patients with hemoptysis had a reduced 6 minute walk distance (356.2 ± 92.5 meters) as compared to patients without hemoptysis (395.1 ± 126.9 meters) ($p = 0.03$). However, other baseline demographic characteristics including diagnosis, complexity of lesion, functional class, and symptoms did not differ among patients with and without hempotysis. Of the 38 patients, 17 had a treatable cause of hemoptysis and received appropriate treatment. The identifiable causes included aorto-pulomanray collaterals, pulmonary thrombosis (2 patients), pulmonary tuberculosis (2 patients), pulmonary artery dissection (1 patient). Treating an identifiable cause reduced the risk of recurrence of hemoptysis by 0.46 (95% CI 0.28–0.64).
Conclusion: Hemoptysis remains a major cause of morbidity in patients with Eisenmenger syndrome. Hemoptysis occurs more frequently in patients with greater exercise limitation. CT pulmonary angiogram immediately following an episode of hemoptysis could identify a potentially treatable cause in nearly half of the patients and such treatment results in lesser recurrence of hemoptysis.

P2515 - RETROSPECTIVE STUDY OF OPTIONAL TREATMENT STRATEGY USING PULMONARY VASOREACTIVITY IN BORDERLINE EIGENMENGER SYNDROME

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Background: This study aimed to evaluate the feasibility, clinical application of pulmonary vasoreactivity testing with inhaled iloprost and to determine the treatment strategy in patients with pulmonary arterial hypertension (PAH) associated with congenital heart diseases (CHD).
Method: This retrospective study was done in single center for 139 cases with CHD diagnosed as PAH through the right heart catheterization from Jan. 2011 to Jul.2016. Inhaled iloprost was delivered during cardiac catheterization by ultrasonic nebulizer via a flow-inflating bag with 0.02 or 0.04 mg. The hemodynamic results and post-catheterization treatment results were reviewed.
Results: The median age of the patients was 32.48 years. The average numbers of systolic pulmonary artery pressure (sPAP) and mean pulmonary artery pressure (mPAP) were 67.39 ± 35.77 mmHg and 44.39 ± 26.30 mmHg. The median pulmonary vascular resistance index was 7.42 Wood units. Among those patients, 58 patients had underwent the pulmonary vasoreactivity test with inhaled iloprost plus oxygen during catheterization (testing group). The average numbers of sPAP and mPAP were 88.32 ± 22.77 mmHg and 57.34 ± 18.37 mmHg in testing group. The median pulmonary vascular resistance index (PVRi) was 10.8 Wood units. 32 (55%) patients were regarded as responder group. The mean PVRi was also significantly decreased in responder group (9.6 to 5.6 vs 12.3 to 11.0 Wood units). After pulmonary vasoreactivity testing, 8 patients had undertaken complete corrective procedure, 19 had received corrective procedure with either fenestration or banding, 4 had undertaken one more procedure/operation followed initial procedure after catheterization, and in only 1 patient, palliative coronary stenting was done. The others were treated with single or combined pulmonary vasodilators without any procedure/operation.
Conclusion: We suggest that even in patients with PAH associated with CHD, pulmonary vasoreactivity test in RHC is essential to determine the treatment strategy of closure or not for borderline Eigenmenger syndrome.

P2517 - LVAD IMMPLANTATION IN DCM CHILDREN

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Objective: To report the usefullness of Excor for DCM children.
Method(s): We have immplanted LVAD (Excor) in 7 DCM children.Age was from 2 month to 12 years old, and body weight was from 2.7 kg to 25 kg. LVAD immplantation was performed using cardiopulmonary bypass under beating heart. 10cc pump were used in 5 children, 15cc in one children, and 30cc in two child. One child has underwent pump exchange from 10cc to 15cc due to body weight gain. INR was controlled from 3.0 to 3.5.
Result(s): 1. Five of 7 children have underwent heart transplantation successfully, one child in Japan, and the other four in the USA. 2. Four of seven children sufferred from infection, and needed antibiotics infusion for a long time. One child sufferring from hemi-paralysis of the leg was free from infection. 3. Children who were immplanted 10cc pump needed 4 times of pump exchange due to pump thrombus during 6 month, and the other 8 times during 11 month.
Conclusion: 1. Excor is a usefull device for DCM children as the bridge to heart transplantation. 2. 10cc pump needed several times of pump exchange due to the thrombus. 3. Fixation of cannula is most immportant to prevent the infection.

P2558 - HEART FAILURE IN CHILDREN B TYPE NATRIURETIC PEPTIDE AS A BIOMARKER FOR INDICATION FOR HEART TRANSPLANTATION AND LONG TERM FOLLOW UP

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Background: Little is known about B-type natriuretic peptide (BNP) in children. The purpose of this study was to evaluate BNP levels profile in children and candidates for heart transplantation.
Patients and Methods: It is a prospective observational study at Heart Institute (InCor) University of Sao Paulo Medical School. The following data was analysed: demographics, ejection fraction (EF), BNP level, clinical outcome (event: transplantation or death). Receiver operating characteristic (ROC) curve were generated to determine the accuracy and optimal threshold of BNP for predicting future outcomes; sample size was calculated for $p < 0.05$; power 80%.
Results: 54 patients were included, 48.1% were male. The majority of subjects had dilated cardiomyopathy (41, 76%) followed by restrictive in 11, hypertrophic in 2. Mean age: 8 years old. Analysis using ROC curve determined the BNP level cut off value of 198.5 pg/ml for predicting future outcomes (specificity: 63.2% and sensitivity: 91.4%).Correlation between BNP level and heart failure (HF) class showed BNP mean level 466.92 pg/ml for HF class I and II and 3004.73 pg/ml for HF class III and IV. ($p = 0.001$). A five- year follow up demonstrated that 48.5% of the patients with HF class I and II were alive and only 3.8% of the cases have had HF class III and IV were free of events.
Conclusions: B-type natriuretic peptide (BNP) could be an additional biomarker and predictor of adverse events in children with heart failure.

P2583 - CARDIAC RESSINCRONIZATION THERAPY FOR TREATMENT OF PEDIATRIC HEART FAILURE AFTER CONGENITAL HEART DISEASE CORRECTION SURGERY

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Cardiac resynchronization therapy (CRT) is a therapeutic option for treatment of severe heart failure (HF) in adults. However, in pediatric patient there's no established consensus. We describe the case of 2 children submitted to CRT for postoperative HF treatment. Patients: ICAB, male, at 18 m, diagnosis of severe dilated cardiomyopathy (LVEF 22%) due to ALCAPA. He has corrective surgery and pacemaker implantation in the 2nd postoperative due to total atrioventricular block, epicardial DDD in LA and LV, but maintained LVEF 20%, QRS > 200 ms and signs of HF (grade IV) with optimized drug therapy. After 1 year, CRT (endocardial electrode implant in RV) was chosen. After 3 months, the patient evolved to grade I HF and maintained good thresholds, QRS <150ms and LVEF 40%. Patient GFG, 5 y, large ASD corrected in 1st year and implantation of pacemaker by TAVB in immediate postoperative period (epicardial VVIR). It evolved with clinical worsening, grade IV HF even with optimized drug therapy ,LVEF 28%, LVBB and QRS > 200ms. Scintigraphy was negative for myocarditis and opted for CRT with multisite electrode on the RA and endocardial RV and new epicardial electrode in LV by lateral thoracotomy, after determination of the best site by strain echocardiogram. After 14 months, he was grade I HF, QRS <150 ms and LVEF 48%. Comments: Therapy of severe HF in the child with CHD has peculiar aspects compared to adults, branch block is common due to extensive surgical scar, as well as associated RV dysfunction. An impairment of ventricular depolarization can lead to mechanical contraction asynchrony, depression of myocardial function and inadequate remodeling. Resynchronization allows simultaneous maximal contraction of both ventricles and improvement of cardiac output. Conclusion: CRT should be considered in cases of postoperative AVB with severe HF, extended QRS and unsatisfactory drug response.

P2597 - CLINICAL OUTCOME OF CARDIOMYOPATHIES DIAGNOSED DURING FETAL LIFE

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Background: There have been a few reports of fetal cardiomyopaties (CMs). However, there is a paucity of information about its assessment, treatment, and outcome.
Methods: We retrospectively reviewed our experience with CMs diagnosed in fetal life from 2008 to 2016. Fetal echocardiographic findings, postnatal laboratory findings, acute phase treatment course, and clinical outcome were investigated.
Results: Five cases (4 male, 1 female) were diagnosed as CMs at mean gestational age of 32 ± 3 weeks. All patients were diagnosed as dilated cardiomyopathy. Left ventricular non-compaction (LVNC) was noted in 4 patients. All patients required mechanical ventilation and continuous catecholamine infusion in the neonatal period. All of them improved clinically to some extent after the acute treatment, though 3 cases died of heart failure at the age of 5, 6, and 9 months. Two patients were discharged home. Both were alive at 6 years' and 15 months' follow-up. Cardiovascular profile scores (CVPS) in fetal echocardiography were 7.5 ± 0.7 points in survivors, and 5.3 ± 1.1 points in non-survivors ($p = 0.04$). Median BNP levels at birth were 210 pg/mL (42-378 pg/mL) in survivors, and 2055 pg/mL (1231-3620 pg/mL) in non-survivors. Mean LV EF was higher in survivors than non-survivors ($33 \pm 4\%$ vs $20 \pm 3\%$, $p < 0.05$). Mean neonatal ventilation period and catecholamine requirement appeared shorter in survivors than non-survivors: 5 ± 3 days vs 29 ± 14 days ($p = 0.04$), 9 ± 2 days vs 35 ± 20 days ($p = 0.07$), respectively.

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Conclusion: The prognosis of fetal CMs was deemed to be poor, as more than half of our cases died in infancy. However, our limited experience suggests the patients with higher CVPS, higher LV EF, and lower BNP have better chance of survival after intensive care in the neonatal period.

P2615 - TREATMENT OF VIRAL MYOCARDITIS

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Aims: Viral myocarditis (VM) is a life treatening disease. Viral and immunohistological mechanism has been implicate in disease progression but etiologic treatment strategies has not demostrated to be effective. Since February 2015 a new protocol has been implemented in our institution wich adds immunosupression or antiviral therapy based on the results of endomyocardial biopsy (EMB). The aim of this study is to describe our experience with the implementation of the new protocol.
Material and Methods: pediatric patients (0-16 years old) admitted in our institution from July 2008 to August 2016 with the diagnosis of VM, were included. We compare the outcome of patients diagnosed before (group A; n = 21) versus after (group B; n = 12) the new protocol. The diagnosis was based upon the presentation of clinical symptoms and the presence of a typical pattern on cardiac magnetic ressonance (CRM) or immunohistological confirmation with EMB. This was only performed in patients with severe disease or without improvement after two weeks of clinical presentation.
Results: median age was 16 months (0 months to 14 years) in group A and 26 months (9 months to 16 years) in group B. Seventeen (81%) versus 8 patients (67%, p = 0,42) needed inotropic support. Mechanical support was needed in 4 (19%) and 3 patients (25%, p = 0,68) in each group. The initial ejection fraction was 35% versus 38% (p 0,42). In group A, 6 patients (28,3%) died or needed heart transplantation. In the group B, the only patient who died (8,3%) suffered a cardiac arrest in another institution and mechanical support was stopped because of neurological damage. Complete recovery was present in nine of eleven patients (81%) in group B.
Conclusions: Immunosupression and antiviral therapy could be useful in VM. This treatment should be based on immunohistological and viral examination of BEM.

P2618 - EXTRACORPOREAL MEMBRANE OXYGENATION IN CHILDREN WITH ACUTE LYMPHOBLASTIC LEUKEMIA

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Background/Hypothesis: Extra Corporeal Membrane Oxygenation (ECMO) is an extreme therapy in critically ill children with respiratory or circulatory failure. Children with Acute Lymphoblastic Leukemia (ALL) have more likely to develop heart failure and respiratory insufficiency, caused by the disease itself or some complication of the treatment. Little is known about the use of ECMO for septic shock or respiratory insufficiency in ALL patients, as this disease can increase the risk of this procedure. There described in the literature only 7 cases of ECMO support for respiratory failure in children with ALL, in two differents centers, in which 5 patients survived until the hospital discharged.
Materials and Methods: We report two cases of ECMO in ALL children.
Results: Case One: A two-year-old female, present with a refractory septic shock due to Moraxella catarrhalis infection and pancytopenia. She was on venoarterial ECMO, and the diagnose of ALL and induction treatment was made during ECMO. The patient was off ECMO after 8 days and is responding well to the ALL treatment as a outpatient. Case two: A two-year-old female, who was newly diagnosed with ALL, had a respiratory failure due to H1N1 influenza and parainfluenza 3 infection after the induction phase of the treatment. She was placed on venovenous (VV) ECMO and received Chemotherapy during ECMO. The patient was of ECMO after 26 days and is recovering from the lung disease progressively and receiving the treatment for ALL. The management of anticoagulation was reviewed because of the bleeding predisposition of ALL. There were no major bleeding.
Conclusions: In our experience, ECMO was feasible and safe in children with ALL. Using very careful monitoring, we can have promising results, bringing hope and life expectancy to theses families and children.

P2620 - MALNUTRITION AS A RISK FACTOR FOR SHORT TERM OUTCOMES AFTER HEART TRANSPLANTATION IN PEDIATRIC PATIENTS AND ADULTS WITH CONGENITAL HEART DISEASE A RETROSPECTIVE ANALYSIS

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Heart failure is an important risk factor for malnutrition, due to cardiac cachexia and high metabolic demand. The outcomes for malnourished adults who have undergone heart transplantation (HTx) have been widely studied, however there are few data on pediatric patients and adults with congenital heart disease. The goal of our study was to analyse the nutritional status of such patients and the impact of malnutrition on the short-term outcomes after HTx. A retrospective review of all children and adults with congenital heart disease submitted to HTx between 2012 and 2015 at our institution was performed. Seventy-four charts were reviewed, with 39 females (52%). The average age was 10 ± 7 years. World Health Organization (WHO) growth-charts were used to analyse the anthropometric data. The average height Z-Score was -1,36 ± 1,97 cm, the average weight Z-Score was -1,83 ± 1,78 and the average body mass index (BMI) Z-Score was -1,17 ± 1,64; Twenty-six patients were diagnosed as malnourished, according to the WHO definition of thinness (BMI Zscore < -2 or < 18 for adults); When comparing the malnourished patients short-term outcomes data with the eutrophic ones, descriptive analysis showed, for the malnourished group, a longer time of orotracheal intubation (6,9 ± 20,4 days versus 2,4 ± 2,8), higher incidence of bacterial infection (19,2% versus 10,4%) and rejection diagnosed by endomyocardial biopsy (57,7% versus 52,1%). The use of renal replacement therapy, the incidence of cytomegalovirus infection and the readmission after first hospital discharge were similar between the groups. Although there are several limitations to our retrospective study, the data above ought to inspire better nutritional evaluation and interventions in the patients awaiting HTx.

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P2625 - PEDIATRIC HEART TRANSPLANTATION IN CHILDREN AND ADULTS WITH CONGENITAL HEART DISEASE STENT IN GRAFT VASCULAR DISEASE – 25 YEARS OF SINGLE CENTER EXPERIENCE

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Background: Heart Transplantation is an option for refractory heart failure and intractable congenital heart disease. Graft vascular disease is one of the most important limitations for long term survival. The aim of this study is to report the use of stent in this situation and the clinical outcome.
Patients and methods: it is a retrospective observacional study in a cohort of children and adults with congenital heart disease submitted for heart transplantation from October 1992 to January 2017 at Heart Institute (InCor) University of Sao Paulo Medical School. The angiogram is routinely performed after transplantation. The demographics and outcome of these patients were evaluated.
Results: 8 of 190 (4.2%) patients could be submitted to stent procedure after transplantation. Mean period between the transplant and the stent procedure was 7.7 years. 4 of them (50%) died. Mean follow-up period of 1.7 years. 3 were re-listed for transplantation.
Conclusions: Stent after heart transplantation is feasible for GVD. However another strategies for treatment of allograft vasculopathy need to be considered such as relisting for transplant.

P2637 - BRIDGING CHILDREN TO HEART TRANSPLANT WITH LEVOSIMEDAN CASE SERIES

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The management of children in the waiting list for Heart Transplant (HTx) is difficult and mortality is high. Once the waiting time may be long, clinical deterioration and cardiogenic shock is a possible outcome. Mechanical circulatory support (MCS) is an alternative to spare time in a scenario where organ availability is scarce, although it's not widely available. Alternatively, levosimendan (LVD) is a vasoactive drug that improves myocardial function. Our goal was to analyse the outcomes of children submitted to HTx after being bridged with LVD. We performed a retrospective review of those children admitted at our institution between 2012-2015.The LVD use consisted of an attack dose of 6-10 mcg/kg/min for 10 minutes, followed by a continuous infusion of 0,1-02 mcg/kg/min for a 24-48 hour period. Eighteen patients were bridged to HTx with LVD during the period. Four patients received both LVD and MCS and were excluded from the analysis. The group bridged exclusively with LVD received a total of 41 cycles. There were 7 females (50%) and the average age was 93,1 ± 68,4 months. Sixty-four percent (9 patients) had acquired heart disease. The average BNP pre-LVD was 2073 ± 2633 and post-LVD was 638 ± 512 mg/dl. The average BNP pre-HTx was 874 ± 781 mg/dl. Regarding the short-term outcomes after HTx the average length-of-stay in the intensive care unit was 51,4 ± 75,2 days and the average duration of mechanical ventilation 2,7 ± 2,9 days. One patient (7%) needed renal replacement therapy, five patients (36%) had seizures after HTx and four patients (28,6%) had infection diagnosed as positive cultures with clinical correspondence. There were no short-term deaths and only one patient died within a 1-year period, due to acute rejection. Although it is an expensive drug, it should be seen as a possibility for effectively bridging children for HTx in a scenario where MCS is not widely available.

P2642 - SPECTRUM OF PERICARDIAL DISEASES IN PEDIATRIC POPULATION PRESENTING TO A TERTIARY CARE CENTRE IN INDIA

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Background: There is a paucity of data on pericardial diseases in pediatric population.
Aim: of the study was to define the spectrum and profile of pericardial diseases in Indian pediatric population presenting to a tertiary care centre. It was an observational study. Children ≤15 yrs who presented to a tertiary care centre were studied from Jan 2010 to Dec 2016. A total of 44 patients were enrolled in the study. The most common pericardial disease entities in descending order were chronic constrictive pericarditis (CCP) 13 (29.5%), pericardial effusion (PE) 13(29.5%), pyopericardium 11(25%), effusive constrictive pericarditis 5 (11.4%) and acute viral pericarditis 2 (4.5%). Tuberculosis was the etiology in 18(40.9%) patients. The spectrum of disease appears to have shifted slightly from chronic constrictive pericarditis to pericardial effusion. Comparison of the etiologies of pericardial effusion (PE) showed a slight increase in frequency of malignancy associated and other causes of PE such as hypothyroidism. The incidence of pericardial calcification is much lesser in pediatric age group one subject in each group (11.1%) as seen in our study. Despite receiving the full course of antitubercular treatment patients with tubercular pleural effusion developed constriction in 60% cases. Patients with purulent pericarditis also progressed to constrictive physiology despite adequate parenteral antibiotics in 45% cases.
Conclusion: In our country Tuberculosis still remains as the most common cause of acquired pericardial disease, despite ATT there is progression to Constrictive physiology in most of the cases.

P2648 - PEDIATRIC HEART TRANSPLANTATION AND OPPORTUNISTIC INFECTION ON NON TYPICAL EPIDEMIOLOGICAL SCENARIOS AWARENESS IS KEY TO SUCCESS

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Background: Hystoplasmosis is the most prevalent mycosis in the United States and its agent is found worldwide, although it is an unusual pathogen in Brazil. Most infections are self-limited, but immunosuppressed patients may develop life-threatening presentations.
Case Report: 15 year-old boy status post-Heart Transplant in 2009 (dilated cardiomyopathy), living in the north region of Brazil, presented to the emergency room in respiratory distress with a history of fever, loss of weight and astenia for 1 month. He had already been treated for pneumonia and cytomegalovirus infection in his hometown, without improvement of the symptoms. His clinical status deteriorated quickly, and he was placed on non-invasive ventilation, broad-spectrum antibiotics (including tuberculosis treatment) and vasoactive drugs whilst investigation was held. Laboratory samples showed bycitopenia (leucopenia and low platelets), Chest X-ray showed diffuse bilateral infiltrate and chest CT revealed bilateral centrilobular nodules. Bone marrow biopsy was negative for infectious pathogens. The Histoplasmosis diagnosis was then carried out through a bronchoalveolar lavage. A 30-

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day course of amphotericin was initiated, with clinical improvement. He was discharged after that period with oral itraconazole.
Conclusion: With the increase in heart transplanted patients, awareness for opportunistic infections should be raised, even if epidemiologically improbable, once targeted treatment is a key for therapeutic success.

P2679 - IS HEART TRANSPLANT FEASIBLE IN CHILDREN WITH RESTRICTIVE CARDIOMYOPATHY WITHOUT PRE TRANSPLANT VENTRICULAR ASSIST DEVICE

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Background: Heart transplant (HTX) in Restrictive Cardiomyopathy (RCM) is a challenge due to the presence of elevated pulmonary resistance. Use of ventricular assist device (VAD) pre-transplant in order to diminish pulmonary resistance is not always possible and increases morbility.
Objective: To describe our experience in HTX with RCM without previous VAD.
Materials and Methods: Retrospective study of our children with RCM receiving HTX from 2006 to 2016. Clinical characteristics, hemodynamic variables pre-post transplant, ischemia time, ECMO need, vasodilator treatment and outcome are described.
Results: From 2006, 89 pediatric HTX were peformed. RCM was Indication in 6 patients (6.7%) with median age of 10 years (4-13) and mean weight: 29.6 Kg (12-66). Etiology: genetic (2), fibroelastosis associated with aortic stenosis (1), Shone syndrome (1) and unknown (2). Hemodynamix variables are shown in the table. All patients had PHT on diagnosis; 4 patients presented PVRI ≥6 Uw/m2, considered high risk patients for HTX. Pulmonary vasodilator medicaments were administered pre transplant. Mean time from diagnosis to listing was 10 months. Waiting list mean time was 2.9 months. Mean ischemia time was 239.5 min (123-310). Pulmonary vasodilator treatment post-transplant: nitric oxid (6), iv sildenafil (1), bosentan (5), oral sildenafil (5) and iloprost (2). All haemodinamic values were normal 1 month after trasplant except in 1 patient whom values are normal after 1 year. 1 patient died in the immediate post-transplant due to cerebral hemmorhage in ECMO. Short term mortality occured in 1 patient because of humoral rejection.
Conclusion: HTX in RCM was possible without previous VAD. Use of pulmonary vasodilators allows listing for HtX with PHT. Decrease in pressure and pulmonary resistance occurs soon after transplant. Choosing a donor with ischemia time ≤240 min to prevent primary graft dysfunction may be a valid strategy to decrease morbimortality.

Table.

	mPAP/ PVRI Dx	PAP /PVRI listing	Treatment PreTx	PAP/PVRI 1 month after TX	ECMO	Treatment PostTX	Follow. up	Outcome
Pat1	27/2,5	25/1.05	Sildenafil	17/0.79	no	NOinh Sildenafil	10 years	Alive
Pat2	38/8.6	42/6.8	Sildenafil Bosentan Milrinone	23/4.1	no	NO inh Sildenafil Bosentan	22 m	Death
Pat3	20/1.6	35/5.6	Sildenafil Bosentan	24/2.8	no	NOinh Sildenafil Bosentan	7 years	Alive
Pat4	32/5.4		Bosentan		Yes	Noinh Sildenafil Bosentan	11 days	Death
Pat5	74/17	48/9.2	Sildenafil Bosentan	30/7	Yes	Iv sildenafil Bosentan NOinh Iloprost	13 month	Alive
Pat6	39/11.8	43/5.4	Sildenafil Bosentan	21/3.6	No	Sildenafil Bosentan NOinh Iloprost	6 month	Alive

mPAP. Pulmonary Artery mean Pressure (mmHg) PVRI : Pulmonay vascular resistance index (Uw*m2)

P2688 - CHARACTERISTICS OF CHILDREN PRESENTING WITH CARDIOMYOPATHY IN AN AFRICAN SETTING – INITIAL FINDINGS OF THE IMHOTEP REGISTRY

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Background: Cardiomyopathy remains a leading cause of morbidity and mortality in children worldwide. Although we see 40 – 50 incident cases annually in Cape Town, the characteristics of childhood cardiomyopathies in African children have not been systematically described. IMHOTEP is the first prospective, open-ended registry of prevalent and incident cases of heart muscle disease in children and adults in Africa. It will expand from pilot sites in Cape Town to other centres in South Africa and subsequently have representation from all 56 countries in Africa to accurately characterize cardiomyopathies on the African continent.
Methods: Commencing August 2016, all incident cases of cardiomyopathy or myocarditis presenting to two referral paediatric cardiac centres were enrolled into a dedicated OpenClinica registry (IMHOTEP African Cardiomyopathy Registry). Details captured include demographics, history, physical examination findings, blood investigation results, genetic screening, chest X-ray, ECG,

Table 1. Patient Demographics

Patient no.	Age at first presentation (months)	Sex	Primary caregiver education	HIV status
1	10	F	High school	NE
2	1.5	M	High school	NE
3	6	F	Primary school	E/neg
4	113	M	Postgraduate	NE
5	2.5	M	Unknown	E/neg
6	32	M	High school	NE
7	3	M	Tertiary	NE
8	108	F	Tertiary	NE
9	5	F	High school	NE
10	3	F	Tertiary	NE
11	89	M	High school	Unknown
12	7.5	M	Primary school	NE
13	0.6	F	Tertiary	NE
14	0.6	F	Tertiary	E/neg
15	0.3	M	High school	E/neg
16	47	F	Tertiary	NE

NE – perinatally non-exposed, E – perinatally exposed, neg – HIV PCR negative

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echocardiogram features and outcome measures (adverse events, hospitalizations, death). Human Research Ethics Committee approvals were obtained prior to implementation of the registry.
Results: Baseline demographic characteristics, modes of initial presentation, echocardiographic parameters and adverse events/outcomes are depicted in Tables 1–4. The majority of patients are infants (11/16, 69%). Four of the 16 patients (25%) are perinatally HIV exposed, but all four tested negative by PCR. The predominant cardiomyopathy phenotype is dilated cardiomyopathy (8/16, 50%) followed by myocarditis (biopsy-proven or clinically suspected, 4/16, 25%). Morbidity and mortality to date has been substantial – two deaths (mortality 13%), average hospitalizations 3/patient, average ICU admissions 1.4/patient, and mean number of days in hospital 30.2 (SD 18).
Conclusions: Already in these early stages, we see the full range of phenotypes represented in African children with cardiomyopathy and a significant mortality and morbidity. The paediatric limb of the IMHOTEP African Cardiomyopathy Registry promises to be a powerful tool to characterize childhood cardiomyopathy in Africa.

Table 2. Mode of initial presentation

Patient no.	Diagnostic category	Modified Ross class	Highest level of care	Days in ICU	Days in hospital
1	Myocarditis	IV	ICU	4	10
2	DCM	III	High care	0	4
3	DCM	IV	ICU	3	7
4	Myocarditis	II	ICU	9	12
5	DCM (due to EAT)	III	ICU	8	24
6	DCM	III	Ward	0	25
7	HCM	II	Ward	0	1
8	DCM	III	ICU	9	27
9	DCM	IV	ICU	3	10
10	DCM	II	Ward	0	8
11	RCM	II	Ward	0	8
12	DCM	III	ICU	11	18
13	LVNC	IV	ICU	7	14
14	LVNC	IV	ICU	9	14
15	Myocarditis	IV	ICU	4	11
16	Myocarditis	I	Ward	0	16

DCM – dilated cardiomyopathy, HCM – hypertrophic cardiomyopathy
RCM – restrictive cardiomyopathy, LVNC – left ventricular non-compaction
EAT – ectopic atrial tachycardia

Table 3. Echocardiographic parameters

Patient no.	LVEDD (mm)	LVEDD (Z score)	LVPWd (Z score)	FS (%)	EF (%)	TAPSE (mm)
1	30	2.6	3.6	12	26	11
2	39	8.5	0.4	9	20	14
3	54	8.2	4.1	10	21	17
4	36	-1.3	1.8	34	66	11
5	30	3.0	0.2	20	34	13
6	45	3.8	2.4	16	30	12
7	21	-2.4	5.9	72	97	21
8	61	7.8	1.9	3	8	8
9	47	16.6	-0.2	12	24	12
10	33	4.5	0.3	6	13	11
11	34	-1.0	-0.8	29	57	13
12	53	16.3	-0.2	8	15	17
13	24	3.9	4.6	20	43	9
14	30	7.9	7.6	16	35	8
15	23	2.5	-1.1	15	33	NM
16	33	0.5	0.3	26	53	13

LVEDD – left ventricular end-diastolic diameter, LVPWd – left ventricular posterior wall diameter at end-diastole, FS – fractional shortening, EF – ejection fraction, TAPSE – tricuspid annular plane systolic excursion, NM – not measured

Table 4. Adverse events and outcomes*

Patient no.	Hospitalizations (total no.)	ICU admissions	Total days in hospital	Alive /died
1	7	3	62	Alive
2	4	3	53	Alive
3	3	2	20	Died in ward
4	1	1	12	Alive
5	2	1	24	Alive
6	1	0	25	Alive
7	3	0	3	Alive
8	3	1	52	Alive
9	3	3	62	Died in ICU
10	5	1	28	Alive
11	4	0	24	Alive
12	2	2	23	Alive
13	4	2	30	Alive
14	3	2	37	Alive
15	1	1	11	Alive
16	2	0	17	Alive

*Current to January 2017

P2701 - HAEMODYNAMIC IMPACT OF PULMONARY VASODILATORS ON FONTAN CIRCULATION

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Aim: The Fontan procedure is the palliative procedure for single ventricle physiology. Pulmonary resistance plays a key role in the success of this operation. There are conflicting data concerning the impact of pulmonary vasodilators on survival and functional capacity among Fontan patients. The aim of this retrospective, single-centre, non-randomised study was to investigate the potential effect of pulmonary vasodilators on pulmonary vasculature in Fontan patients.
Method: Twenty-eight patients with single ventricle physiology were enrolled in the study. Eighteen patients were treated with pulmonary vasodilators: nine patients after Glenn procedure (GroupA) and nine patients after Fontan completion (GroupB). Ten patients with single ventricle after Glenn procedure were enrolled as a control group (GroupC). The primary end point was

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to assess changes in haemodynamic profile and the diameter of the pulmonary branches after two right heart catheterisations. Adverse events were recorded.
Results: Mean age ± SD was 3.2 ± 1.5 years (Group A), 26.8 ± 12.7 years (Group B), and 3.0 ± 1.0 years (GroupC). Patients included in Group A had smaller pulmonary arteries (Nakata index: 158 ± 50 vs 212 ± 68 mm2/m2; p = 0.047) and reduced arterial compliance (34.3 ± 15.4 vs 56.3 ± 25.6 mm2/[m2*mmHg]; p = 0.03) at baseline compared with Group C. After treatment, Nakata index and pulmonary compliance increased in patients treated with pulmonary vasodilators (Group A), while remaining stable in the control group (Nakata index: +23 ± 21% vs −7 ± 16%, p = 0.003; pulmonary compliance +80 ± 49% vs −8 ± 34%, p = 0.001). Similar results were found in Group B (Nakata index: pre-168.6 ± 70.7 mm2/m2; post-204.9 ± 97.5 mm2/m2; p = 0.026).
Conclusions: Pulmonary vasodilators reduce pulmonary artery resistance and increase vascular compliance, pulmonary artery diameter and cardiac output in Fontan patients. Therefore, pulmonary vasodilators may be used before the Fontan procedure in patients who are at high risk of Fontan procedure failure.

P2718 - BETA 1 ADRENERGIC RECEPTOR ANTIBODIES IN DILATED CARDIOMYOPATHY IN CHILDREN AND THEIR RESPONSE TO AN IMMUNOADSORPTION THERAPY

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Background: β1-adrenergic autoantibodies have been reported in sera from adult patients with dilated cardiomyopathy (DCM). Their removal have demonstrated a hemodynamic benefit.
Hypothesis: Determine the presence of β1-adrenergic autoantibodies in children with DCM and the potential hemodynamic benefit upon their removal by immunoadsorption (IA).
Material and Methods: We tested β1-adrenergic autoantibodies in 17 children The etiology of DCM was : genetics (4), myocarditis (3), mother lupus-associated congenital heart block (3), and idiopathic (7). All patients evidenced ejection fraction (EF) lower than 45%. Immunoadsorption cycle (4 sessions per cycle) therapy was conducted in patients with positive β1-adrenergic autoantibodies. BNP was determinated before and after IA cycle.
Results: Antibody testing resulted to be positive in 7 patients.IA therapy was performed in all of them. The technique was well tolerated. BNP concentration was lowered after each IA cycle in all patients. 3 patients with highest levels of autoantibodies showed the poorest clinical outcome and were transplanted. 2 patients remained negative after 1 cycle of IA and they recovered their heart function completely. 1 patient improved after several cyles of IA (BNP decrease and FE from 30% to 45%) but levels of antibodies remained positive. He died due to sepsis 15 months later. 1 patient with recurrent myocarditis due to parvovirus with an increase in levels of antibodies in each episode was treated with immunoglobulin and IA. Currently, after 5 years.

P2721 - INITIAL EXPERIENCE IN CHILDREN WITH THE USE OF MACITENTAN IN PULMONARY ARTERIAL HYPERTENSION

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Background: Macitentan is an orally active and dual endothelin receptor antagonist (ERA) whose efficacy was assessed in pulmonary arterial hypertension (PAH) on adult patients.It has a slow receptor dissociation kinetic and more affinity with lipophilic membranes compared to other ERA, reducing the risk of hepatoxicity.
Materials: We report our initial experience of its use in two pediatric patients with PAH.
Results: CASE 1: A 2-year-old child who was diagnosed with severe PAH with coincidental atrial septal defect and patent ductus arteriosus. At cardiac catheterization, the mean pulmonary arterial pressure (PAPm) was 70 mmHg, pulmonary arterial wegde pressure (PAWP) 5 mmHg, and pulmonary vascular resistance (PVR) 22.5 UW·m2. She was treated first combining sildenafil with bosentan, and then with ambrisentan, but both had to be discontinued as a result of a significant increase of liver enzyme levels. At the age of 6 years, weighting 23 kg, treatment with macitentan was associated to sildenafil, at a dose of 5 mg/day, without side effects.
CASE 2: A 15-month-old premature infant who was diagnosed after birth with severe valvular aortic stenosis and borderline left ventricle. A Norwood-Sano procedure was perfomed at 6-month-old. The postoperative period was complicated with persistent hypoxemia, and haemodynamic criteria for PAH was diagnosed (PAPm 31 mmHg, PAWP 10 mmHg, PVR 4 UW·m2). He was treated with sildenafil and bosentan, but bosentan had to be discontinued because of hepatoxicity. At the age of 15 months, weighting 7,7 kg, he started treatment with macitentan, at a dose of 2,5 mg/day, without side effects. After 6 months, a second palliative step (Glenn procedure) was performed, without postoperative complications.
Conclusions: macitentan is a promising drug in PAH treatment in the pediatric population because of its low risk of hepatoxicity. Multicentric studies are necessary to determine safety, effectiveness and dosage in children.

P2736 - OXIDATIVE STRESS IN HIGH ALTITUDE IN PATIENTS WITH TRISOMY 21 IS REVERSIBLE BY VITAMIN C

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Introduction: Due to the underlying genetic disposition patients with Trisomy 21 are prone for increased oxidative stress. This becomes a significant problem when environmental hypoxemia is present (i.e. congenital cardiac surgery respiratory tract infections, high altitude exposure).
Methods: To evaluate the influence of high altitude hypoxemia we investigated children with Trisomy 21 without congenital heart disease who are living permanently in high altitude in La Paz, Bolivia (3800-4100 m above sea level) and compared them with age and sex matched control. In a second step we treated both groups with Vitamin C.
Results: 33 children with Trisomy 21 (TR21) and 58 controls, age 7.5 ± 5.4years vs. 7.6 ± 4.8years (Tr21 vs. controls), saturation in room air 88.5 ± 3.8% vs. 88.9 ± 2.8% (TR21 vs. controls). RV/RA gradients have been higher in TR 21 patients at baseline (33.5 ± 10.1 vs. 20.2 ± 6, p <0.00001) as did Hb (15.2 ± 2.2 vs. 14.1 ± 1.1, P <0.01). After treatment with Vitamin C for 1 months RV/PA gradient significantly decreased in the TR21

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group (31.7 ± 5.5 to 27.5 ± 6.9 mm Hg, P < 0.01) while in controls no effect could be observed (23 ± 4.8 vs. 25.5 ± 4.3). Oxygen saturation remained unchanged.
Conclusion: Exaggerated hypoxic pulmonary hypertension in Trisomy 21 patients without congenital heart defect appears to be triggered by oxidative stress since one month administration of oral Vitamin C could induce a significant decrease in RV/PA gradients while healthy control did not show any positive effect under Vitamin C treatment. This beneficial effect was not related to any improvement of arterial oxygen saturation.

P2744 - RECURRENT HYPERCAPNIA AND HYPOXEMIA IN CONGENITAL CENTRAL HYPOVENTILATION SYNDROME (UNDINE SYNDROME) LIKELY ACTS AS ENDOTHELIAL PRECONDITIONING

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Introduction: Undine syndrome (US) is a rare disease with severely impaired central autonomic control of breathing and dysfunction of the autonomous nervous system. The incidence is estimated to be at 1 of 200,000 livebirths. Due to reccurent hypercapnia and hypoxia we hypothesized that these patients have higher risk for pulmonary arterial hypertension (PHT) and longterm systemic vascular dysfunction. We examined 7 patients with US at baseline and high altitude in regards to pulmonary artery pressure and systemic vascular disease and compared them with 6 age and sex matched subjects.
Methods: 14 patients with US (18.6 ± 4.1 y) and 15 age and sex matched healthy controls (17.9 ± 1.5 y) were examined at 550 m and at 3883 m above sea level with echocardiography (to measure pulmonary artery pressure). Vascular function was assessed by pulse wave velocity (PWV) and carotid intima-media thickness (IMT).
Results: US patients had mildly elevated RV/PA gradients at 550 m above sea level while no relevant RV/PA gradient was found in the controls (25.8 ± 8.0 vs. 20 ± 4.3 mmHg, P = 0.03). In 3883 m above sea level all US subjects showed only mild increase in RV/PA gradients similar to the controls (33.5 ± 17.8 mmHg vs.29 ± 21 mmHg). Systemic vascular function was impaired in US subjects as shown by an increased PWV (8.0 ± 1.2 vs. 7.0 ± 0.9 m/s, P = 0.02, US vs. control) and IMT (418.4 ± 34.2 vs. 367.7 ± 50.6 μm, P < 0.01, US vs. control).
Conclusion: US patients show impaired systemic vascular function. Despite this findings Undine patients do not show evidence for marked pulmonary hypertension during rapid ascent to high altitude. We speculate that endothelial preconditioning due to intermittent hypoxemia might have a protective effect.

P2797 - RISK OF CELLULAR OR ANTIBODIES MEDIATED REJECTION POST HEART TRANSPLANT IN CHILDREN EFFECT OF ECMO OR VAD USE

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Background: Studies suggest ventricular assistance device (VAD) or ECMO may increase HLA sensitization in children with heart transplantation (HT) but it is difficult because of confounding factors. We have studied the episodes of cellular rejection or antibodies mediated rejection (AMR) attributable to ECMO or VAD use.
Methods and Materials: We reviewed retrospectively the data of all children <18 years of age transplanted in our center between December-2008 and January-2017. Patients on cardiac support (CS) prior and after the transplant were compared to children never on CS.
Results: A total of 37 children were transplanted, 14 (37,8%) on CS prior at HT, 8(21,6%) after HT (2 with CS preHT). Mean age was 7.35 years (5 m -17,5y); twelve children (32%) had congenital heart disease (previous surgeries). Types of CS were: ECMO in 7 patients (35%), ECMO and Berlin Heart(BH) in 8(40%), VAD or BH 4 and 1 VAD + BH. Mean time of cardiac support was 53,2 days (8 hr- 330 d). Similar immunosuppression protocol was used, except induction: Basiliximab in 17(46%) and thymoglobulin in 20(54%). Basiliximab was related with more cellular rejection (p = 0,049). Cross match was negative in all cases. Only 1 patient had PRA > 10% prior at HT. Episodes of cellular rejection or AMR in patients with/without CS were similar (0 = 0,330/0,693). There was not statistical association with type or durability of CS, diagnosis or previous surgeries. Only one patient died at 11 days related with AMR episode (PRA +). Median time of follow-up was 3,5 year. Overall actuarial survival after cardiac transplantation was 94,5% at 1 month, 89% at 1 year, and 86% at 5 years. Freedom from rejection and survival was similar in our patients with or without CS.
Conclusions: The use of VAD or ECMO in our patients prior or after the HT is not associated with a higher incidence of rejection post-transplant. Overall survival was similar in our patients with/withour CS.

P2820 - PERICARDITIS IN CHILDREN A SINGLE CENTER EXPERIENCE

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Background: Pericarditis is an inflammation of the pericardium and often occurs with effusion. It may be infectious or noninfectious, secondary to a systemic disease. The aim of this study was to analyze the clinical findings, course and treatment of pericarditis in children treated at our center.
Material and Methods: Retrospective analysis of all children admitted in a pediatric cardiology unit with pericarditis between 2003 and 2015. Patient characteristics were summarized using frequencies and percentages for categorical variables and medians with percentiles for continuous variables.
Results: Fifty patients were analyzed. The median age was 14 years (interquartile range 9 to 15 years) and 40 patients (80%) were male. The most common presentation form was acute pericarditis (80%). Thirty-five patients (70%) presented with chest pain and 26% reported fever. Cardiomegaly was identified in 11 patients' (22%) chest x-ray, sixty percent had an abnormal ECG and 44 patients (80%) had transthoracic echocardiogram changes, namely pericardial effusion. In 17 cases (34%) there was myocardial involvement with raised cardiac biomarkers. Forty-eight percent of patients presented with infectious pericarditis and the pathologic agent was identified in half of these cases, however no seasonal

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pattern was noticed. Postpericardiotomy syndrome diagnosis was made in 5 cases. The first-line therapy was acetylsalicylic acid in half of patients. Pericardiocentesis was performed in 12 patients. The median length of stay was 9 days (interquartile range 3 to 11 days). There was symptom recurrence in 7 children.
Conclusions: Pediatric pericarditis is a rare condition with scarce epidemiological data available. In this study, infectious acute pericarditis was the most common presentation and about one third of patients had also myocarditis. The symptom recurrence rate is not negligible and is probably related with the type of therapy employed. Recent proposed guideline changes may potentially improve this incidence.

P2824 - MICROFLUIDIC CHIP BASED QUANTIFICATION OF CIRCULATING ENDOTHELIAL CELLS IN PATIENTS WITH PULMONARY ARTERIAL HYPERTENSION

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Background: Circulating endothelial cells (CECs) have been discussed as potential biomarkers in patients with pulmonary arterial hypertension (PAH). Current protocols for isolation and quantification of these cells are laborious and time-consuming. The aim of this study was to develop and apply a microfluidic chip capable of enumerating CECs (CD146 +) from human whole blood in patients with PAH and age matched controls.
Methods: EDTA blood was collected and injected into polymeric microfluidic chips containing microcolumns pre-coated with anti-CD146 antibody. Captured cells were immunofluorescently stained for additional stem and endothelial cell markers.
Results: The CEC capture chip was first validated against conventional flow cytometry ($r = 0.89$). In a cohort of 66 patients with three forms of PAH (idiopathic/heritable, drug-induced, and connective tissue disease), CEC numbers were significantly increased by 3-5-fold in PAH subjects vs. matched controls. CEC numbers were comparable across PAH sub-classes and were not related to age, postmenopausal status, or body mass index.
Discussion: The CEC capture chip requires 400 μL of whole blood and may serve as bedside test for the screening and monitoring of patients with PAH and other diseases related to vascular injury.

P2832 - IMPACT OF HEART FAILURE CLINIC IN PEDIATRIC AGE

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Objectives: It has been reported that heart failure clinic (HFC) can improve the outcome of adult heart failure patients, in terms of reduction of rehospitalisation, adherence to therapy and improvement in quality of life. The aim of current study was to determine whether a specific HFC is effective also in a paediatric population.
Methods: Since 2013, a specific HFC including two cardiologists and 1 dedicated nurse has been developed in our Department. A multidisciplinary approach and specific disease protocols have been applied. Children affected by: 1) left ventricular ejection fraction <45% in case of dilated cardiomyopathy or after surgery, 2) restrictive and 3) hypertrophic cardiomyopathy 4) Fontan failure and 5) pulmonary hypertension have been referred to our HFC. To evaluate the impact on mortality, rehospitalisation, analysis of comorbidities and compliance to therapy, we made a comparative analysis on 291 children followed between 2011 and 2015, before and after the specific program.
Results: Our population (11.2 + 5 yrs old; 52% male) was represented by: 45% patients affected by cardiomyopathies, 50% CHD with ventricular dysfunction or Fontan failure. After the introduction of this program, a significant reduction of new urgent rehospitalization was observed (2011 vs 2014 75% vs 33%, $p < 0.001$; 2011 vs 2015 75% vs 12%, $p < 0.001$; 2011 vs 2016 75% vs 14%, $p < 0.001$). Acute HF admissions were significantly reduced over the time (2011 vs 2016 52% vs 13%, $p < 0.001$). No differences in terms of mortality have been observed. Major reasons for rehospitalisation were: respiratory (52%), gastroenterological infection (33%), arrhythmias (12%). Major comorbidities included: nutritional aspects, respiratory disease and orthopaedic problems and 45% of patients had more than two comorbidities.
Conclusions: in our center, HFC allowed to significantly reduce the rehospitalisation rate in a paediatric population.

P2839 - INTRACLASS CORRELATION BETWEEN ECHOCARDIOGRAPHIC FINDINGS AND HEMODYNAMIC DATA IN PATIENTS DIAGNOSED WITH IDIOPATHIC PULMONARY HYPERTENSION IN FUNDACION CARDIOINFANTIL – INSTITUTO DE CARDIOLOGIA IN BOGOTA COLOMBIA

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Objectives: Pulmonary hypertension (PH) is defined as a mean pulmonary arterial pressure ≥ 25 mmHg. The diagnosis of idiopathic PH is made after a thorough study clinical and paraclinical including complete anatomical and functional assessment. The aim of this study is to describe echocardiographic findings and hemodynamic data obtained by cardiac catheterization and assess their degree of correlation.
Materials and Methods: Descriptive cross-sectional study and analysis of matching variables through the comparison of pulmonary pressure measured by echocardiography and catheterization through Interclase correlation coefficient (ICC) and the Bland Altman. The study population were children aged 0 days to 18 years who presented Fundación Cardioinfantil – Instituto de Cardiología in Bogotá (Colombia) diagnosed with idiopathic PH evaluated between January 2010 and December 2013.
Results: 29 patients diagnosed with idiopathic PH studied with echocardiography and cardiac catheterization were included. The degree of PH documented by echocardiography corresponded to severe in 38% of patients. After cardiac catheterization, the degree of PH was classified as severe in 66% of cases. In 18 patients (62%) pulmonary vascular reactivity test was performed, finding positive response in 7 cases (39%). The concordance analysis of pulmonary pressure measured by echocardiography and catheterization was performed in 20 subjects, with a confidence level of 95%. When

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comparing the measurement results in both tests is a difference of 15 mmHg which is statistically significant. The tests achieved an ICC of 0.55, with 95% of 0.16 to 0.79, which represents a force of moderate agreement.
Conclusions: In the present study the severity of PH corresponded to severe in most patients evaluated for cardiac catheterization. Performing pulmonary vascular reactivity test for both diagnosis and accurate diagnostic decisions is essential. The concordance analysis of pulmonary pressure measured by echocardiography and catheterization through Intraclase Correlation coefficient and the Bland Altman documents a force of moderate agreement.

P2840 - THREE DECADES OF PEDIATRIC HEART TRANSPLANTATION DATA FROM THE SPANISH HEART TRANSPLANTATION REGISTRY

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Introduction: Heart transplantation (HT) represents the treatment of advanced refractory heart failure in children. The Spanish Heart Transplantation Registry collects clinical data of pediatric HT since May 1984.
Objective: Describe the clinical characteristics and outcome of pediatric CT in our country.
Material and Methods: We analyzed the clinical characteristics of donors and recipients and the short and long-term outcome in children under 16 between 1984 and 2014.
Results: A total of 391 HT were performed, 237 males (60%) with a mean age of 6.9 ± 5 years. 19 patients (15.5%) had some degree of renal dysfunction. The main cause were cardiomyopathy in 214 (57.5%) and congenital heart disease in 145 (39%). 182 (50.7%) children were transplanted in urgent status and 48 (26%) had a circulatory assist device (CAD), 45 were implanted in the last decade. The most common cause of death in donors was traumatic brain injury. The mean follow-up time was 6.3 ± 6.4 years. The most frequent complications were rejection (48%), infections (52%), graft vascular disease (10.7%) and tumors (4.7%). The leading cause of death was graft dysfunction followed by acute rejection and sudden death. Survival at first month was 82.2%, one year 74.1%, 5 years 68%, 10 years 59.2% and 15 years 51.4%.
Conclusions: Short and long term outcomes in Spain are consistent with those reported in the world literature, including a continuing trend towards better survival over the years. At present, 1 in 4 children who are transplanted in urgent status has CAD. It would be necessary to analyze the impact of the increasing use of CAD on perioperative survival and during the first year of transplantation as well as to identify risk factors for complications.

P2862 - REDUCED INFLUENCE OF TGFβ SIGNALING OVER BMP SIGNALING IN THE MCT + AC SHUNT RAT MODEL OF PULMONARY ARTERIAL HYPERTENSION

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Background: Pulmonary arterial hypertension (PAH) is a cardiopulmonary disorder in which cellular proliferation in the arterioles is responsible for excessive proliferation of the arteriolar cells obstructs the vasculature, which increases pulmonary arterial pressures. As important regulators, endothelial cells are anticipated to be the major culprits in PAH. Interestingly, the presumed dysbalanced TGFβ/BMP pathway has never been dissected on endothelial level in rat models for PAH.
Aim: To map the expression and regulation of TGFβ and BMP pathway mediators to assess the presence and nature of their dysbalance in rat models for PAH.
Methods: The pathway components Tgfbr1, Bmpr2, Smad2, Smad8, Serpine1 and Id3 were assessed by RT-qPCR, and pSMAD2/3, pSMAD1/5/8, PAI-1 and ID-3 were assessed with IF in three different PAH models, i.e the MCT + AC-shunt, MCT only and AC-shunt only at early (d14) and late (d28) stage.
Results: IF staining of intra-acinar arteries showed no differences in localization or expression of the proteins pSMAD2/3, pSMAD1/5/8, PAI-1 and ID-3. In isolated pulmononary ECs, gene expression of all pathway components in all models except the Flow only model was downregulated in compliance with the PAH progression (table1). The ΔCT value ratio of Tgfbr1:Bmpr2 is 1:2.8 in the Sham group versus 1:1.2 on MCT + Flow day 28. This is mimicked in the MCT only group, where the Tgfbr1:Bmpr2 ratio dropped from 1:2.8 to 1:1 on day 28. The Flow only model shows an initial drop of 1:1.6 on day 14, but corrects to 1:2.3 on day 28.
Conclusion: In isolated endothelial cells of the MCT + Flow and MCT only PAH models, the influence of TGFβ pathway over the BMPR2 pathway gradually decreases. In the Flow only model this aspect ratio normalizes. This suggests that TGFβ inhibiting agents could have no effect in rat models of PAH, and might also not benefit patients with CHD-PAH.

Table 1. Gene expression downregulation versus Sham

	Tgfbrl	*Bmpr2*	*Smad2*	*Smad8*	*Serpinel*	*Id3*
MF14	1.1	2.0	0.9	2.2	0.6	1.4
MF28	6.9	26.4	7.6	20.0	2.8	5.8
M14	0.9	2.7	1.1	2.3	1.7	1.5
M28	3.3	22.3	4.2	41.5	1.8	4.5
F14	1.1	2.8	1.5	1.9	1.2	1.6
F28	1.4	1.6	1.3	1.4	1.6	1.1

P2869 - LATE "FAILING" FONTAN IN A TERTIARY CENTER IN PORTUGAL 18 YEARS FOLLOW UP

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Background: The Fontan procedure represents the final surgical stage of the troublesome univentricular pathway. We aimed to describe some of the factors associated with "Late Failing" Fontan (FF) circulation.
Materials and Methods: Clinical medical records of children/young adult born after 1987 submitted to Fontan procedure and followed up at a tertiary centre in Portugal, were reviewed. Patients with evidence of early FF were excluded. Late FF was defined as: death/

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transplant, Fontan conversion, heart failure (HF) symptoms, protein-losing enteropathy (PLE) or plastic bronchitis.
Results: Fourty-six patients were included, 25 males (54,3%), with 4-29 yo at present, with complex CHD (table 1). Thirty patients had single LV anatomy, 12 single RV anatomy and four biventricular heart. Total cavopulmonary connection (TCPC) was performed in all patients and the median age was 5,7yo (P25-P75: 4,4-7,6yo). Forty-one patients had an extracardiac conduit (EC) and five a lateral tunnel (LT); 37 were fenestrated Fontan. Eleven patients (23.9%) developed late FF: five PLE, five HF and one death. No patient were submitted to cardiac transplant or Fontan conversion. The only deceased patient was 13yo (5,2yo after surgery) with malignant arrhythmia. The median time of FF presentation was 5.0yo(P25-75: 0.6-9.9)after the Fontan surgery. Comparing with the non-FF, FF patients more frequently had LT Fontan (4/5 vs EC conduit 7/41, $p < 0,001$) and arrhythmias (4/8 vs no arrhythmia 5/30, p 0.049). All patients with ventricular dysfunction (5/46) developed FF ($p < 0,001$). No other significant differences were found between the groups (ventricular morphology, size of pulmonary arteries, presence of fenestration, collateral vessels).
Conclusions: Less than 25% of our patients developed late FF. The lack of associations between some of the studied factors and FF might be due to the small sample. Future studies on the factors predicting the outcome of Fontan procedure are imperative to improve the management of these patients.

Table 1. Baseline Characteristics of all patients, Non-"Failling" Fontan and "Failling' Fontan Patients.

	All n = 46	Non-"Failling" Fontan n = 35	"Failling" Fontan n = 11	*p*
Demography				
Age at analysis (years)	15.0 (P25-75: 11.8-18.0)	14.2 (P25-75:9.3-23.5)	16.5 (IC 95%:14.0-17.3)	0.570
Male sex	25 (54.3%)	22 (62.9%)	3 (27.3%)	0.039
Age at Glenn Surgery	2.0 (P25-75: 0.8-4.2)	2.0 (IC 95%: 0.8-4.7)	2.6 (IC 95%: 1.0-4.2)	0.887
Age at Fontan Surgery	5.7 (P25-75: 4.4-7.6)	5.9 (IC 95%: 4.5-7.8)	4.6 (IC 95%: 4.1-7.5)	0.157
Congenital Heart Disease				0.058
Double-inlet LV	11	(23,9%)	7 (20.0%)	4 (36.4%)
Tricuspid atresia	10	(21,7%)	9 (25.7%)	1 (9.1%)
Pulmonary atresia	9 (19,6%)	8 (22.9%)	1 (9.1%)	
Hypoplastic Left Heart	9 (19,6%)	8 (22.9%)	1 (9.1%)	
Syndrome				
Double-outlet RV	3	(6,5%)	1 (2.9%)	2 (18.2%)
L-TGA	2	(4,3%)	2 (5.7%)	0 (0%)
D-TGA	1	(2,2%)	0 (0%)	1 (9.1%)
Unbalanced AVSD	1	(2,2%)	0 (0%)	1 (2.2%)
Type of Total Cavopulmonary Connection				0.002
Extracardiac conduit (EC)	41 (89%)	34 (97.1%)	7 (63.6%)	
Lateral tunnel (LT)	5	(11 %)	1 (2.9%)	4 (36.4%)
Fenestration	37 (82.2%)	27 (77.1%)	10 (100%)	0.095
Other characteristics				
Hypoplastic Pulmonary arteries	10/40 (25.0%)	7 (22.6%)	3 (33.3%)	0.512
Ventricle Dysfunction	5 (11%)	0 (0%)	5 (100%)	< 0.001
Presence of Collateral vessels	10/29 (34.5%)	7 (35.0%)	3 (33.3%)	0.930
Dysrhythmias	8/38 (21.1%)	4 (13.8%)	4 (44.4%)	0.049

P2910 - TEN YEAR EXPERIENCE OF A PEDIATRIC HEART TRANSPLANT PROGRAM IN BRASILIA BRAZIL

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Introduction: Heart transplantation is the last remaining therapeutic option for children with end-stage heart disease. Building a pediatric heart transplantation program is still challenging in developing countries.
Objectives: To describe the early and midterm results of a pediatric heart transplantation program in Brasilia.
Methods: We included all pediatric patients (≤18 years) who were listed for a heart transplantation in a pediatric cardiac center between 2007 and 2016.Parameters analyzed were obtained prospectively over the years.
Results: 42 children with a mean age of 5.7 years (2 months to 15.5 years) were listed for cardiac transplantation as intention to treat. Cardiomyopathy was present on 35 children(83%) and 23 (55%) were aged ≤5 years. 32 children (76%) were on INTERMACS ≤3, while 21 (50%) were on mechanical ventilation. 17 children (41%) died while on waiting list and 3 children were delisted (clinical improvement). Mortality in the waiting list was higher in children aged <1 year (86% x 36%) and those who were in INTERMACS ≤3 (50% x 10%). Cardiac transplantation was done in 22 children (52%), with a waiting list time of 94 days (2 to 502 days). Mean cold ischemia time was 140 minutes (57 to 270 minutes). ICU stay after transplantation was 18 days (3-48 days) and hospital stay was 43 days (3-84 days). Mean follow-up time was 3.2 years (35 days at 7.2 years), and survival rate at 30 days, 1 and 5 years were 86%, 82% and 82%, respectively. At least one episode of rejection occurred in 63% of the patients, and one child had a lymphoproliferative disease after 2 years of transplantation.
Conclusions: Early and midterm results obtained with this new pediatric cardiac transplantation program in Brasilia were quite satisfactory although mortality rate on waiting list was very high especially in children less than 1 year of age.

P2912 - DESCRIPTIVE STUDY ON ANTHRACYCLINE INDUCED CARDIOTOXICITY IN PAEDIATRIC ONCOLOGY PATIENTS IN SINGAPORE

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Introduction: Anthracyclines as chemotherapeutic agents are widely used to treat childhood cancers. Cardiotocixity is a known adverse effect of anthracycline treatment; however, its incidence in Singapore is unknown. The aim of this study was to evaluate the epidemiology of paediatric anthracycline-induced cardiotoxicity in Singapore.
Method: This was a single-center, retrospective analysis of 25 paediatric oncology patients who were treated with anthracyclines and subsequently developed cardiotoxicity. Primary data analysed were quantified echocardiographic findings, details of anthracyclines used, onset of cardiotoxicity and treatment, as well as their outcomes. Secondary points included patient profiling and patient follow up.
Results: Doxorubicin was the most commonly used anthracycline, 52% of patients underwent doxorubicin treatment with a mean cumulative dose of 206 mg/m2. Other anthracycline treatments used were daunorubicin, epirubicin and idarubicin with a mean doxorubicin-equivalent cumulative dose of 217 mg/m2. The cohort developed cardiotoxicity at 10.8 years with decreased ventricular function being the most common diagnosis. The entire cohort had their cardiotoxicity diagnosed by imaging (mean fractional shortening: 22%, mean left ventricular end-systolic dimension: 45.5mm, mean left ventricular end-systolic dimension: 36.1mm), with 52% being diagnosed as an inpatient. The cohort's

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cardiac disease free time was 19.2 months and had a total follow up period of 3.8 years with a 68% survival rate (65% of whom remained in remission). The number of cardiac events was 2.71 and 20% of the cohort developed congestive heart failure. The most common treatment used were angiotensin converting enzyme inhibitors.
Conclusion: This was the first local study of its kind, with the aim to gain a deeper understanding of the local paediatric population's cardiac response to anthracyclines, which would help in future management, monitoring and counselling of such patients, and pave the way for more related studies.

P2975 - CLINICAL DETERMINANTS OF OPTIMAL BETA BLOCKER DOSE IN CHILDHOOD HYPERTROPHIC CARDIOMYOPATHY

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Background: Pharmacokinetic studies of carvedilol have established that infants require 4.3 times higher dose/kg body weight to obtain comparable plasma-levels to adults, and 2-11 year-olds 2.9 times, and 12-15 year olds 1.4 times higher. Propranolol is metabolized by the same enzyme, and accordingly it is to be expected that physiologically equivalent beta-blockade effect would require different propranolol-doses across this age range.
Methods: Consecutively diagnosed patients (age range 0-19 yrs) with hypertrophic cardiomyopathy (HCM), n = 105, between 1983-2000 at the John Radcliffe Hospital, Oxford, UK, and between 2000-2015 at the Queen Silvia Children's Hospital, Gothenburg, Sweden, that were treated with propranolol (or equivalent doses metoprolol or bisoprolol) because of symptoms, outflow-tract obstruction at rest, or progressively worsening hypertrophy. The doses were titrated up to achieve a profound beta-receptor blockade as judged by 24 h Holter and heart-rate response to exercise testing according to standardized protocol (Östman-Smith et al, JACC,199;34:1813-22). Beta-blocker dose required were analyzed in four age-ranges, and correlation analysis with different clinical features was performed.
Results: Beta-blocker dose correlated significantly with age at diagnosis ($p < 0.0001$, correlation coefficient (cc) -0.411), septum-to cavity ratio ($p < 0.0001$; cc = 0.606), Detroit Z-score septum ($p < 0.0001$; cc = 0.540), left-atrium-to-aortic ratio ($p = 0.0007$; cc = 0.351) and left ventricular outflow-obstruction ($p = 0.0004$; cc = 0.390). Propranolol doses/day required for optimal beta-blockade at different age ranges were 0-1.9yrs (n = 35): median 11.6 mg/kg [IQR 7.2-17.4]; 2-6.9yrs (n = 16): 6.5 [5.2-9.4] mg/kg; 7-12.9yrs (n = 20): 7.4 [5.8-10.5] mg/kg; 13-19.9yrs (n = 34): 6.4 [4.6-8.0] mg/kg. In the whole group HCM-related to Noonan-spectrum disorders was significantly correlated to a higher dose, but this was an age-related effect, as correlation did not remain significant when patients were compared within the same age range.
Conclusions: Infants require substantially higher doses/kg body weight for adequate beta-blockade than older children. Above the age of two, degree of hypertrophy, outflow tract obstruction and indicators of diastolic dysfunction also influence dose-requirement significantly.

P2997 - NEUROVASCULAR COMPLICATIONS IN CHILDREN SUPPORTED ON BERLIN HEART EXCOR DEVICE EARLY RISK FACTORS FOR STROKE

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Introduction: The incidence of neurovascular complications (NC) after Berlin Heart (BH) EXCOR ventricular assist device (VAD) remains high despite considerable VAD experience and aggressive management of anticoagulation.
Aims: To identify perimplantation risk factors for neurovascular complications after BH VAD implantation.
Methods: Retrospective cohort study of children supported with BH VAD at our institution from November 2004 to May 2015. Pre-, peri-, post-implantation clinical data, neurovascular complications, coagulation profile, platelet function tests and inflammatory markers were analysed. NC were identified by head CT and/or brain MRI performed as clinically indicated.
Results: A total of 75 children were supported with BH VAD for end-stage heart failure at a median age 2.9 years (1.2-8.8 years) and median weight 13 kg (9-26 kg). The most common indication was for end-stage cardiomyopathy (70/75). The median time on VAD support was 39 days (16-102 days). Overall survival to transplant or recovery was 85% (64/75). NC occurred in 21/75 (28%) at a median duration 15 days (8-35 days) after implantation. Of the children with NC, 11 had an ischaemic stroke, 7 had a haemorrhagic stroke, and 3 had subdural haematomas. After NC, 7/21 (33.3%) died due to devastating neurological complication, and out of the remaining 14, 12 were transplanted and 2 recovered. Using multivariable analysis, total blood products given within 24 hours after VAD implantation [HR 1.04 (95% CI 1.02-1.06)], weight (per 10 kg) [HR 0.65 (95% CI 0.56-0.75)], and postop C-reactive protein levels [1.01 (95% CI 1.00-1.01)] were independently associated with NC.
Conclusions: Risk of NC remains high and these events usually occur early after BH VAD implantation. Postoperative blood product requirement and CRP potentially identify modifiable risk factors for stroke.

P3005 - A STUDY SHOWING THE EFFICACY OF NO FOR PULMONARY VASULCAR BED REACTIVITY DELIVERY WITH NOVEL IMPROVISED HOOD

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Context- Study of delivery of NO for diagnostic cardiac catheterization in paediatric patient BIPAP vs paediatric Hud.
Aims and Objective: Effective delivery of NO with the HUD which is comparable to BIPAP delivery, in pediatric spontaneously breathing patient with the use of NOXBOX machine.
Methods and Material: This is comparative prospective pilot study, to assess effectiveness of delivery of NO via HUD, compared to that of BIPAP mask. Study population is grouped into two based on mode of delivery as BIPAP receiving group and HUD receiving group. Each group contain 5 spontaneously breathing pediatric patient with underlying pulmonary hypertension, and, NO is delivered to assess reversibility of pulmonary vasculature.

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Result: data collected as above, in HUD group, we were able to deliver NO effectively in acceptable range in all 5 patients same in BIPAP group.
Conclusion: We found that this system of NO delivery by HUD is a simple delivery system with wide applicability. We have shown that it has similar efficacy as that of BIPAP to deliver NO and O2. Its safety profile appears to be comparable to that of standard, more complicated, delivery systems.

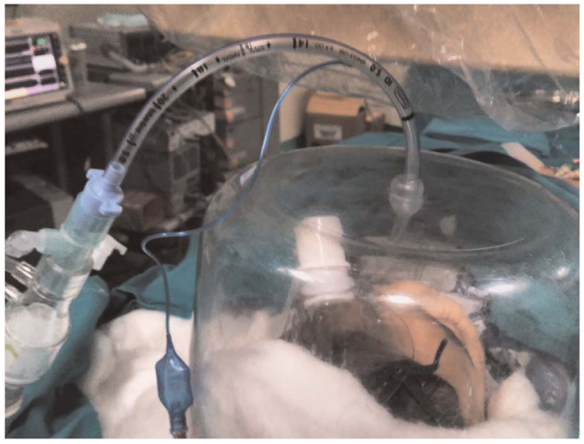

Figure.

P3007 - PERCUTANEOUS IMPELLA DEVICE FOR MECHANICAL CIRCULATORY SUPPORT IN CARDIOGENIC SHOCK PEDIATRIC EXPERIENCE

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Background: Management of acute cardiogenic shock frequently involves use of aggressive pharmacologic support. Patients who fail to respond to standard medical therapies might require mechanical circulatory support. Recently, Impella ventricular assist device has been approved for management of cardiogenic shock in adults. Experience with Impella support in children is limited. We present our experience with Impella device in a pediatric cardiovascular intensive care unit (CVICU) at a freestanding children's hospital.
Methods: All patients treated with Impella device between September 2014 and June 2015 were included in retrospective analysis. Demographic, hemodynamic and laboratory data were reviewed and reported using descriptive statistics. Data was compared using Student's t-test with significance level at $p < 0.05$.
Results: Total of 13 Impella devices were implanted in 10 patients: CP (n = 6), and 5.0 (n = 7). Median patient age was 18 years (13,35), weight of 77 kg (56,124), and BSA of 1.97 m2 (1.61,2.47). Average length of CP support was 4 days (median 3.5 (1,10)) and 5.0 device 19 days (median 12(10,45)). Average flow for CP and 5.0 was 3.0 L/min and 4.2 L/min. There was a significant improvement in PCWP post implantation ($p = 0.03$) and 24 h vasoactive score ($p = 0.03$). Compared to immediate post device implant, the lactate at 24 hours and creatinine at 5 days were significantly lower ($p < 0.05$). Median LDH level was 1813 U/L (791,7264) and plasma hemoglobin <30 mg/dL (<30,86) after 5 days of support. Insertion site bleeding occurred with 6 devices (46%). All patients survived to ICU discharge.
Conclusion: Impella support for acute cardiogenic shock shows hemodynamic benefits with a limited morbidity and no early mortality in pediatric CVICU. This modality could be considered as a support option in management of older children with acute cardiogenic shock to achieve end organ recovery or as a bridge to clinical decision.

P3013 - TWELVE YEARS EXPERIENCE WITH PEDIATRIC CARDIOMYOPATHY IDENTIFYING OUTCOME RISK FACTORS

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Back Ground: Pediatric cardiomyopathy (CM) is a rare but serious and often life-threatening condition. In children, cardiomyopathy is often a part of multisystem disorder, which requires the attention of multiple subspecialists.
Objective: The aim of this work is to describe the most common demographic, clinical features and its relationship to the outcomes of children diagnosed with cardiomyopathy in the clinic during that period.
Materials and Methods: This retrospective descriptive study was carried out at the Pediatric cardiomyopathy Clinic, children's Hospital, Cairo University, to review the files of patients diagnosed with cardiomyopathy in the period from 2004 to 2016.
Results: The cases diagnosed with cardiomyopathy during that period were 1282 cases. The patient files included 948 cases with DCM, 260 cases with HCM, 45 cases with RCM, 29 cases with LVNC and one case with RVD. The age of the patients ranged from 10 days to 18 years with a mean age at presentation of 3.2 years. The most common clinical presentation was dyspnea (84.9%) followed by heart failure while chest infection were (50.6%). A family history was positive in (48.5%). Treatment was mainly treatment of heart failure or LVOTO. 142 (11.1%) of our patient died and 396(39.6%) with lost follow up more than 5 years. There were significant risk factors for mortality including reduced shortening fraction, ejection fraction, repeated intensive care unit admission and history of a metabolic disorder. Insignificant risk factors will be also identified and described.
Conclusion: The current study demonstrates that children with CM are at particular risk for death during the initial ICU admission and reduced FS/LVEF may be predictive of outcome, Patients with significant risk factors may benefit from closer outpatient observation and perhaps, earlier consideration for stem cell therapy and cardiac transplantation.

P3024 - THE CORRELATIONS BETWEEN THE GENOTYPE OF FBN1 MUTATION AND PHENOTYPE IN 85 UNRELATED TAIWANESE PATIENTS WITH MARFAN SYNDROME

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Objectives: To explore the correlations between FBN1 genotype and phenotype in Taiwanese Marfan syndrome.
Methods: From May 2007 to Sep 2013, a total of 125 patients (mean age: 25.1 ± 14.6 years, 58% male) diagnosed as Marfan syndrome fulfilling the Ghent criteria and confirmed with FBN1

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gene mutation were enrolled. Mutation was identified by denaturing high-performance liquid chromatography or High-resolution melting analysis.
Results: A total of 85 mutations were identified in 125 proved MFS patients. The mutation comprised 60 missense, 9 nonsense, 15 frame shift, and 1 whole exon deletion. Most mutations located in exon 11-24 (28.2%), then exon 25-40 (22.4%), and in cbEGF like domain (63.5%), then TGFBP domain (18.8%). The distributions of age and young patients number were even in the whole gene without statistical significance. The correlation of genotype and phenotype based on the exonic location showed that mutations in exon 1-24 were associated with major ocular symptoms. Mutations in exon 41-57 were associated with major cardiovascular and skeletal symptoms but less ocular involvement. Mutations in exon 58-65 were associated with minor symptoms in three systems. Based on the classification of domain, mutations in NH2 region were associated with major cardiovascular and ocular symptoms. Mutations in COOH region were associated with minor symptoms. Mutations in hybrid domain were associated with major cardiovascular symptoms. Based on mutation type, nonsense mutations were associated with major cardiovascular symptoms.
Conclusions: Genotype information is essential for diagnosis of Marfan syndrome. The usefulness of predictive genetic testing in FBN1 mutations requires further investigation.

P3028 - THROMBOELASTOGRAPHY PLATELET MAPPING IN CHILDREN SUPPORTED WITH THE BERLIN HEART EXCOR PEDIATRIC WHEN MIGHT IT NOT WORK AND WHAT ARE THE ALTERNATIVES

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Background: Pediatric centers rely primarily on Thromboelastography and Platelet Mapping™ (TEG/PM) to dose-adjust platelet inhibitors in children supported with the Berlin Heart EXCOR Pediatric. It is unclear whether TEG/PM™ results are accurate because PM has not been validated in children. We sought to evaluate the agreement between 3 platelet function assays in a Berlin Heart recipient who had a poor PM™ response to high-dose platelet inhibitor therapy.
Methods: Platelet function was measured serially using TEG/PM, standard optical platelet aggregometry (OA) and Multiplate (MP) in a 3 year-old boy supported with a Berlin Heart. For TEG/PM, ASA response was evaluated by % inhibition to Arachadonic Acid (AA) while DPA and clopidogrel response was evaluated by % inhibition to adenosine di-phosphate (ADP). Platelet aggregation for OA was evaluated by % aggregation and for MP by standardized units (out of 100).
Results: Following EXCOR implant, ASA was uptitrated to a target dose of 30 mg/kg/day, DPA to 4 mg/kg/day and clopidogrel to 1 mg/kg/day as per institutional protocol. At target doses, the %AA inhibition from TEG/PM (N = 34) ranged from 0% to 33% and the %ADP inhibition ranged from 0% to 35% consistent with a poor response. By contrast, OA revealed a %AA and %ADP aggregation of of 5-35% and MP revealed ASA and ADP aggregation of <30 U, both consistent with good platelet response.
Conclusion: In selected patients PM has poor agreement with OA raising questions about its validity. By contrast OA has good agreement with PM, which requires a smaller blood sample (<1 cc) suggesting it may have advantages over PM. Further studies are need to determine the optimal method for measuring platelet function in children.

P3035 - SILDENAFIL THERAPY OF PULMONARY HYPERTENSION IN CHILDREN LONG TERM (10 YEARS) SURVIVAL OF PATIENTS

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Background: Determination of the efficacy, safety and tolerability of specific drugs for the treatment of pulmonary arterial hypertension in children is a crucial task in pediatrics. Results of a prospective study (2004-2016) estimating long-term (over 5 years) survival of patients received sildenafil monotherapy are interesting for the choice of treatment both by patient and the doctor.
Methods: Clinical examination, cardiopulmonary test (bicycle ergometer), hemodynamic evaluation, chest X-ray, BORG dyspnoea scale, ophthalmologic (including colour vision) evaluation, electrocardiography, ECHO-cardiography, clinical labs, BNP, pro-BNP were performed.
Results: Four patients with primary pulmonary hypertension (WHO class II-III), and 9 patients with pulmonary hypertension associated with congenital heart disease (class II-III) were followed. 5-year survival in primary pulmonary hypertension was 100%, in pulmonary hypertension associated with congenital heart disease - 89%. Three patients with congenital heart disease have underwent successful surgery 3, 8 and 9 years after start of the therapy (previously considered as inoperable). Four patients died: 20 months (ventricular fibrillation), 6 years (sudden cardiac death), 7 years (perforated duodenal ulcer) and 7 years (progression of pulmonary hypertension amid pulmonary infectious diseases) after start of the therapy.
Conclusions: Sildenafil monotherapy in patients with primary pulmonary arterial hypertension and associated with congenital heart disease pulmonary arterial hypertension improves long-term survival.

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