Volume 29 • Supplement 1 • Pages S1–S196
doi:10.1017/S1047951119000489
Cardiology in the Young
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53rd Annual Meeting of the Association for European Paediatric and Congenital Cardiology (AEPC)

Fibes–Conference and Exhibition Centre, Seville, Spain | May 15–18, 2019

YIA-1

Administration of steroid could be associated with larger coronary artery aneurysm in patients with Kawasaki disease – a single center experience

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Background: The number of patients with giant coronary artery aneurysm (CAA) has not been expectedly deceased so much according to the 24th the national survey for KD in Japan.
Objective: To elucidate the risk profiles of KD patients who develop CAA with special reference to steroid treatment.
Design/Methods: We performed a retrospective analysis of patients who were evaluated for KD between July 2003 and July 2018. Demographics of the patients were obtained from the medical records and characteristics of the CAA were evaluated by using echocardiogram and coronary angiography, which included number of CAA, the location of CAA (bilateral, left only, right only), the size of CAA (Z score), the length of CAA (1 segment, 2 segments only, and over 3 segments). We divided the patients into 2 groups who used steroid or not and compared these characteristics. Furthermore, we compared the the incidence of complication of CAA and cardiac events between these 2 groups.
Results: A total of 29 patients were suspected to have CAA by echocardiogram and coronary angiography was performed in our institution during the study period (24 males, median age 24 months (range; 2 months -7years). These KD patients were treated by aspirin and intravenous immunoglobulin (IVIG) in 17 cases (60%, non-steroid group), aspirin and IVIG plus steroid in 11 (37%, steroid group) cases, and unknown in 1 (3%) case. There were no significant differences in the number of CAA, the location of CAA, the length of CAA between steroid group and non-steroid group. But the size of CAA was significantly larger in patients with steroid group for Z score (steroid 8.7 vs non-steroid 6.3, p=0.02). During the median follow up period of 33 months (range: 1 month - 10 years), we observed 12 (41%) cases of coronary artery stenosis, 5 (17%) cases of coronary artery occlusion, of whom 2 cases required coronary artery bypass grafting.
Conclusions: CAA size was larger in KD patients who used steroid than that of non-users. However, cardiac complication of CAA and cardiac events were comparable between these groups. Further prospective, multicenter studies are needed to confirm these findings.

YIA-2

Can platelets facilitate adhesion of Staphylococcus aureus to cardiac graft tissue, used in RVOT revalvulation, and lead to increased risk of infective endocarditis (IE)?

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Introduction and Objectives: RVOT reconstruction in congenital heart disease can be surgically done using cryopreserved pulmonary homograft (CH) and alternatively xenografts such as the bovine jugular vein (BJV) valved conduit. Despite this good therapeutic alternative recent clinical studies report an increased risk of IE in BJV. This raises the question of why such valves are more prone to IE than homografts. We investigate whether different tissues promote interactions with blood components and therefore enhance the risk for S. aureus adhesion to valve tissue.
Methods: Grafts were incubated with fluorescently labeled plasma fibrinogen (Fg). Then, S. aureus adhesion to the tissues was assessed under flow conditions using a flow chamber system after tissue preincubation with human plasma, albumin or serum. Moreover, tissue susceptibility for platelet interaction was evaluated upon blood perfusion using a colorimetric assay. To document a contribution of Fg-mediated pathway to the interplay bacteria-tissue-platelets, bacterial mutants and anti-platelet drugs were employed. Fg binding to tissues was quantified with fluorescence microscopy and bacterial adhesion was evaluated by CFU counting on blood agar. Bacteria and platelets were visualized on the tissues with confocal or electron microscopy.
Results: Bovine pericardium presented higher protein binding ($P < 0.05$) compared to BJV and CH. Although not significant,

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

there is a trend towards higher Fg interaction with BJV than CH. After incubation with plasma S. aureus adhesion to BJV increased significantly under flow compared to control conditions (serum $P < 0.05$ and albumin $P < 0.001$). Both bacterial and platelet adhesions to BJV were greater in relation to CH ($P < 0.01$). Moreover, deletion of clfA hampered bacterial adhesion to BJV ($P<0,05$) as well as eptifibatide significantly reduced ($P < 0.001$) platelet reactivity towards BJV.
Conclusions: Our results indicate that the role of Fg-mediated pathway is important for both bacterial and platelet recruitment to endovascular tissues. The grafts differ in susceptibility to bind platelets what might promote bacterial adhesion, where the interaction Fg-integrin αIIbβ3 receptor takes a part. Future studies will focus on endothelialization of grafted valves and how this affects lesion formation and development of infection. Moreover, antiplatelet treatment will be addressed to study its effect on bacterial recruitment.

YIA-3
Is it true that the one and a half ventricle repair strategy has poor prognosis? - postoperative hemodynamic assessment compared with the Fontan procedure

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Introduction: The one and a half ventricle repair (1.5 VR) strategy has been used for patients with a hypoplastic or dysfunctional right ventricle. To date, the 1.5 VR strategy is considered inferior to the Fontan procedure. Most studies assessed heterogeneous populations, and hemodynamic advantages and disadvantages of these strategies remain unclear. We aimed to clarify hemodynamic characteristics of 1.5 VR strategy using one-to-one pair matching of patients who underwent either procedure.
Methods: We retrospectively reviewed patients who underwent 1.5 VR with simultaneous catheterization and magnetic resonance imaging (MRI) between July 2009 and August 2018. Anatomical diagnosis-, age-, and sex-matched patients who underwent Fontan procedure were analyzed via pair matching. Post-operative hemodynamics and flow analysis parameters were examined on phase-contrast MRI.
Results: Overall, 15 patients (7 with corrected transposition of the great arteries and 4 who displayed pulmonary atresia with intact ventricular septum) were evaluated in each group. Median follow-up after either procedure was 9.0 years. The inferior vena cava (IVC) pressures were lower (median, 6.0 vs. 10.0 mmHg; $p = 0.01$), superior vena cava(SVC) pressures were higher (median, 11.0 vs. 9.0 mmHg; $p = 0.09$), respectively, in the post-1.5 VR group than in the post-Fontan group. The IVC and SVC blood flows were lower in the post-1.5 VR group than in post-Fontan group (median, 1.5 vs. 1.8 L/min/m2; $p = 0.01$; median, 0.9 vs. 1.1 L/min/m2; $p = 0.003$, respectively). Systemic and pulmonary blood flows in the post-1.5 VR group were lower than in post-Fontan group (median, 2.4 vs. 2.8 L/min/m2; $p = 0.01$; median, 2.7 vs 3.0 L/min/m2; $p = 0.04$, respectively). The IVC pressure in the post-1.5 VR group gradually increased over time, but remained high from the early postoperative year in the post-Fontan group. Of 7 patients who underwent ultrasonography, no post-1.5 VR patient had finding of hepatic congestion, whereas 3 post-Fontan patients had congestion.
Conclusions: Lower IVC pressures observed in the post-1.5 VR group could be associated with abdominal organ disorder alleviation. All blood flow parameters were lower in the post-1.5 VR group than in post-Fontan group; however, whether this is an advantage remains uncertain.

YIA-4
iCMR Evaluation of Single Ventricle Patients: A Pilot Study

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Introduction (or Basis or Objectives): We describe our early institutional experience performing real-time interventional CMR (iCMR) procedures to evaluate the Fontan circulation using the MRI compatible wire (angle-tip Emeryglide MRWire, Nano4Imaging, Aachen, Germany) to guide catheters for a right heart catheterization (RHC), left heart catheterization (LHC), and Fontan fenestration test occlusion (FFTO) when indicated. The FFTO procedure is traditionally performed in the cardiac catheterization laboratory to evaluate patient's candidacy for potential Fontan fenestration device closure (FFDC).
Methods: Patients underwent an iCMR procedure using a dilute gadolinium-filled balloon-tip catheter in combination with the MRWire for Fontan pathway/RHC, LHC, and FFTO under real-time MRI visualization. A recently developed passive catheter tracking technique (real-time spoilt gradient echo (TFE), FA 35-45 degrees, TE 1.3ms; TR 2.7ms; 40 degrees partial saturation (pSAT) pre-pulse) was used to visualize the gadolinium-filled balloon, MRWire, and cardiac structures simultaneously. MRWire visualization is enabled due to distal markers creating susceptibility artifact.
Results: MRWire was used on 14 out of 20 single ventricle (SV) patients undergoing iCMR. Median age and weight were 5.6yrs and 17.7kg, respectively (range: 2-16yrs and 11.5-43.6kg). SV anatomy (n = 20): 11 pre-Fontan evaluations, 8 post-Fontan patients for PLE/cyanosis evaluations (6 fenestrated and 2 non-fenestrated), and 1 pre-Glenn evaluation.
Real-time MRI-guided RHC (n = 14), LHC/aortic pull back (n = 14), and FFTO (n = 2) was successfully performed in all patients when the MRWire was used. No complications were encountered. Time taken for first pass RHC, LHC/aortic pull

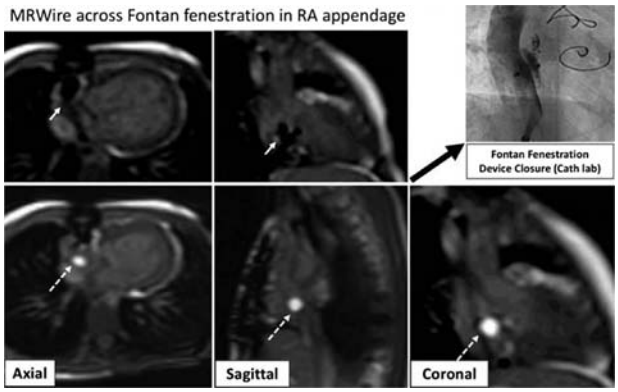

Figure 1.
*Successful FFTO followed by successful FFDC in the Cath Lab **White Arrow** – MRWire; **Dashed White Arrow** – Gadolinium-filled ballon*

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

back, and FFTO was 4.9, 2.9, and 6.5 minutes, respectively. Patients were transferred to the fluoroscopy lab if further intervention was required including FFDC, balloon angioplasty, and/or coiling of collaterals when indicated.
Conclusions: Feasibility for diagnostic RHC, LHC, and FFTO iCMR procedures with the MRWire in SV pediatric patients is demonstrated. Novel real-time TFE with optimized FA-pSAT has facilitated simultaneous visualization of the catheter balloon tip, MRWire, and cardiac/vessel anatomy during iCMR procedures. These cases describe a more thorough evaluation of Fontan pressures and cardiac output before FFDC by using accurate flow, ventricular volumes, and cardiac output measurements from real-time MRI with simultaneous catheter based pressure measurements.

YIA-5
The outcome of patients with functional single ventricle after pacemaker implantation—what makes it poor and what can we do?

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Introduction: In the patients with functional single ventricle, it is known that the risks of death and several kinds of complications are high, both before and after the Fontan procedure. Permanent pacemaker implantation for bradyarrhythmia is one of the factors related with poor outcome. However, the detailed mechanism is not fully recognized.
Methods: A retrospective chart review of patients with single ventricle who have undergone permanent pacemaker implantation at Fukuoka Children's Hospital was performed. The patients were categorized into three groups, according to the existence of a ventricular lead and the frequency of ventricular pacing: Group A, 11 patients with pacemaker with atrial lead only; Group B, 12 patients with ventricular lead and ventricular pacing rate <50%; Group C, 15 patients with ventricular lead and ventricular pacing rate >50%. Group C was subsequently divided into two subgroups, according to the location of the ventricular lead, as the 7 patients with apical ventricular pacing lead and 8 patients with non-apical ventricular pacing lead for further analysis. In all cases, the pacing leads were epicardial leads.
Results: Groups A and B did not have any mortality, whereas group C had survival rates of 58.9% and 39.3% after 10 and 20 years, respectively, of pacemaker implantation. Among the post-Fontan patients, there was no difference among the three groups in terms of ejection fraction and proportion of atrioventricular valvular regurgitation. The BNP (brain natriuretic peptide) significantly increased according to the frequency of ventricular pacing at 11.7, 20.3, and 28.4 pg/ml for groups A, B, and C ($p = 0.04$). In Group C, the outcomes did not show significant difference between two subgroups. However, the BNP was significantly lower in post-Fontan patients with ventricular apical pacing lead than in those with non-apical pacing lead (27.0 pg/ml vs. 82.8 pg/ml, $p = 0.03$).
Conclusions: A higher frequency of ventricular pacing was related with poor outcome and higher BNP, probably implying the association with ventricular dyssynchrony. The apex was the optimal site of an epicardial ventricular lead for patients with functional single ventricle.

YIA-6
Evaluation of Fontan failure using the severity of FALD as secondary organ disease

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Introduction: Despite improved survival, Fontan palliated patients are prone to failure of the Fontan circulation and second organ dysfunction in the form of Fontan-associated liver disease (FALD). In this study we established a graduation of FALD based on laboratory and ultrasonographic liver assessment and analyzed its applicability for definition of Fontan failure.
Methods: Liver assessment was performed in 90 consecutive patients with a median age of 17.3 years [IQR 14.9]. The extend of FALD was graded as mild, moderate and severe due to laboratory parameters (liver enzymes, Fibrotest®), liver ultrasound and liver stiffness measurement (Fibroscan®). Hemodynamic assessment was performed using echocardiography and cardiac catheterization. Fontan failure was defined as active protein-losing enteropathy, impaired cardiopulmonary exercise capacity measured by spiroergometry with a VO2max below 45% of the age adjusted standard value or more than 2 hospitalisation periods due to cardiopulmonary decompensation within 12 months.
Results: FALD was graded as mild, moderate and severe in 46, 34 and 10 patients (51.1%; 37.8%; 11.1%). The extend of FALD significantly correlated with exercise capacity ($p<0.001$) and systolic ventricular function based on echocardiography ($p=0.003$). Invasive pressure measurement revealed a strong correlation between the extend of FALD and Fontan pressure ($p<0.001$), mean pulmonary artery pressure ($p=0.002$) and end-diastolic ventricular pressure ($p=0.003$). Fontan failure was detected in 17 patients (18.9%) and correlated with severity of FALD ($p<0.001$). Death occurred in 4 patients with failing Fontan circulation, FALD was graded moderate in 1 and severe in 3 patients ($p=0.004$).
Conclusion: Detection of FALD is mandatory for Fontan surveillance and monitoring of the failing Fontan. Severity of FALD significantly correlates with exercise capacity, ventricular function and hemodynamics of Fontan palliated patients. FALD graduation might be a useful diagnostic for definition of Fontan failure and evaluation for heart transplant.

O1-1
Predictors of Appropriate Interventions in Children with Hypertrophic Cardiomyopathy and a Primary Prevention Defibrillator: An international multi-center study

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Introduction: Primary prevention (PP) ICD are increasingly placed in children with Hypertrophic Cardiomyopathy (HCM) to prevent SCD using adult criteria. The reliability of PPICD to prevent SCD is unclear.
Methods: We collected data on HCM children ≤20 years with PPICD. Risk factors (RF) for SCD were 1) family history (FH) of SCD; 2) syncope; 3) maximal Left ventricular (LV) wall thickness (LVWT) Z value > 5; 4) non-sustained VT (NSVT) on ambulatory ECG; and 5) abnormal BP response to exercise (ABPR).
Results: Of 347 patients (age 14.61± 4.18), with PP-ICD, appropriate interventions (AI) occurred in 55 (16%). RF presence pre-ICD implant was FH-SCD in 114 (33%); LVWT in 230 (66%); NSVT in 25 (7%); ABPR in 105 (30%) and syncope in 84 (20%). Follow up after ICD implant was 0.07 to 32.95 (mean 8.82 ± 6.03) years and incidence of AI was 18.40 cases per 1000 person-years. AI incidence for each RF was: FH-SCD: 20/114 (17.5%); LVWT 36/230 (15.7%); NSVT 4/25 (16.0%); ABPR 18/105 (17.1%) and syncope 22/84 (26.2%). Risk of AI based on # of RF were: 5/31 (16%) for 0 RF; 16/126 (13%) for 1 RF; 20/146 (14%) for 2 RF; 12/36 (33%) for 3 RF and 2/8 (25%) for 4 RF.
Conclusions: The incidence of AI in HCM children with PP-ICD based on adult criteria was high. Syncope was the commonest RF associated with AI. All other RF's had similar incidence in patients with AI. Presence of ≥ 3 RF was associated with AI.

O1-2
Mechanical Synchrony And Contraction Efficiency In Left Ventricular Apical Pacing In Children: Comparison To Normal Controls

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Introduction: Left ventricular apical pacing (LVAP) has been reported to preserve LV function in chronically paced children with complete atrio-ventricular block (CAVB). We sought to evaluate the long-term effect of LVAP on LV mechanics as compared normal controls.
Methods: 36 paediatric patients with CAVB and LVAP in absence (N=22, group A) or presence of repaired structural heart disease (N=14, group B, systemic LV in all) and 25 age-matched normal controls (group C) were retrospectively studied. Echocardiography was performed after a median of 2.9 (IQR 1.9-6.2) years of pacing. LV function and synchrony were evaluated by M-mode, 2D echocardiography and speckle tracking analysis in standard apical 4-chamber (4CH), 2-chamber (2CH) and parasternal short axis (at the level of mitral valve) views. Data were analyzed using the ECHOPAC and MATLAB software.
Results: See table for LV function and mechanical dyssynchrony data. LV contraction inefficiency measured by systolic stretch fraction (proportion of myocardial stretch and shortening during systole) was higher in groups A and B than C in both the 4CH and 2CH views ($p<0.001$ and $=0.035$, resp.) and correlated significantly with the apical to basal mechanical delay ($p=0.001$ for both). There was no correlation between any of the dyssynchrony parameters and LVEF.

Group	LVEF [%]	GLS [%]	SPWMD [ms]	Apical to basal delay [ms]	Septal to lateral delay [ms]	Antero-septal to posterior delay [ms]	Anterior to inferior delay [ms]
A	68 (63-70)	−20.4 (3.1)	13 (28)	67 (18-87)	−1 (−10−6)	1 (−7−13)	0 (−5−6)
B	61 (56−69)	−20.1 (2.7)	13 (25)	82 (43-128)	0 (−3−12)	7 (−8−15)	6 (−8−35)
C	60 (56-61)	−21.0 (2.4)	47 (22)	19 (2−39)	3 (−1−12)	4 (−1−24)	13 (1−24)
p	=0.004	=0.723	<0.001	<0.001	=0.419	=0.692	=0.044

Median (IQR) or mean (SD). GLS=global longitudinal strain. LVEF=left ventricular ejection fraction. SPWMD=septal to posterior wall motion delay.

Conclusions: LVAP maintains mechanical synchrony between LV septum and free wall at the price of a significant apical to basal mechanical delay associated with LV contraction inefficiency as compared to healthy controls. Global LV systolic function is, however, not negatively affected. Results are similar in both presence and absence of structural heart disease.

O1-3
Extracardiac conduit Fontan procedure has a similar arrhythmogenicity to classic Fontan procedure at long term follow-up of patients with univentricular physiology without heterotaxia syndrome

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Introduction: Arrhythmias are a common comorbidity in subjects who underwent Fontan procedure (FP). Many studies reported that the extra-cardiac conduit (ECC) FP have a lower arrhythmia burden, as compared to the atriopulmonary FP. However, the ECC cohorts described in the literature have a significantly shorter follow-up (FU) than the atriopulmonary FP cohorts. Our aim was to investigate the prevalence and time of onset of arrhythmic complications in patients who underwent ECC with long-term FU at a single institution.
Patients and methods: We retrospectively evaluated 348 patients who underwent ECC at our institution (1987-2013). Patients with heterotaxy were excluded for their intrinsic arrhythmic risk; other exclusion criteria were previous non-ECC FP, ECC take-down

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

and loss to FU (overall 119 patients). Seven patients (3%) died during FU (only 1 sudden death). The remaining 222 patients (60.4% males) had a maximum FU after ECC of 30 years. Mean age at FU was 19.2 years, while at surgery - 4 years. Native cardiovascular anomalies were mainly univentricular hearts of left ventricular type (60%).
Results: Four patients experienced complete atrioventricular block before surgery, thus postoperative arrhythmic FU was performed on 218 subjects. Overall arrhythmia burden was 28.5% (62 patients): 4.6% early postoperative arrhythmias and 23.8% late postoperative arrhythmias, with a mean length of time (δt) from ECC to development of arrhythmia of 9 years. All the early-onset arrhythmias were bradyarrhythmias. Among the late-onset arrhythmias 21.5% (47 patients) were bradyarrhythmias (δt from ECC 8 years), 13.5% (29 patients) were tachyarrhythmias (δt from ECC: 11.5 years for supraventricular tachycardias and 13.5 years for ventricular tachyarrhythmias). Fourteen (6.4%) patients had both a bradi- and a tachy-arrhythmia. 28 subjects (12.8%) required permanent pacemaker implantation. No implantable cardioverter-defibrillators were placed. The incidence of arrhythmia increased with age – mean age at FU of the arrhythmia group being 22 years compared with 18.2 years in the no arrhythmia group (p=0.0026).
Conclusions: This study demonstrates that arrhythmias are a frequent complication also in patients with ECC Fontan without heterotaxy, suggesting that the length of time from surgery, rather then the type of FP, is the main risk factor for development of arrhythmia.

O1-4
High ECG risk-scores correlate with late Gadolinium enhancement on magnetic resonance imaging in HCM in the young

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Introduction: Hypertrophic cardiomyopathy (HCM) is the most common inherited cardiac disease. HCM is characterized by gradual thickening of the myocardium and disturbed myocardial composition. ECG risk-score >5p has been shown to detect pediatric patients with subsequent cardiac arrest with high sensitivity. Myocardial fibrosis measured by cardiac magnetic resonance (CMR) has also been proposed as a risk-factor for cardiac events. Previous studies have shown disturbed myocardial perfusion surrounding the fibrotic areas. CMR is time consuming and expensive. We sought to assess whether ECG analysed by ECG risk-score could be used as an indicator of fibrosis or perfusion deficit in HCM as measured by CMR.
Methods: A single center cohort of 50 individuals; 41 patients from 29 family-pedigrees, HCM-patients (n=24), age 7-31 years, 17 individuals at risk of HCM (phenotype-negative mutation-carriers,n=8; first-degree HCM-relative,n=9), and 9 healthy controls, underwent CMR and 12-lead ECG. CMR was performed using late Gadolinium enhancement (LGE) and Adenosine stress-test to identify fibrosis and myocardial perfusion defect. ECG was analyzed according to the ECG risk-score method (Eur Heart J 2010;31:439), and categorical data compared with Fisher's exact test.
Results: The majority of HCM-patients presented <19 years of age. ECG risk-score >5p was significantly more common in the HCM group, median 3p [IQR 0-9p], vs. controls and individuals at risk of HCM; median 0p [0-1p], (p=0.001); and significantly more common also compared to individuals at risk of HCM (p=0.005). In patients with positive LGE median risk-score was 8p [3-10p]. An ECG risk-score in the high-risk range (6-14p) correlated with positive LGE on CMR (p<0.001), with specificity 97%, sensitivity 57%, positive predictive value (PPV) 89% and negative predictive value (NPV) 85%. Including perfusion defect in analysis did not increase sensitivity, and specificity and PPV remained unchanged.
Conclusions: Myocardial fibrosis on CMR (LGE) correlate with high ECG risk-scores. ECG risk-score in this study is very specific and quite sensitive. Since ECG is easily performed and available in almost every healthcare facility, ECG risk-score allows you to prioritize your high risk patients and could be an inexpensive complementary tool in risk stratification of HCM in the young.

O1-5
Cardiac Sympathetic Denervation As Adjuvant Treatment To Prevent Malignant Arrhythmias In Pediatric Population

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Background: Cardiac sympathetic denervation (CSD) is a second-line treatment for adrenaline-sensitive channelopathies refractory to drugs.
Objective: Present our series of pediatric CSD for LQTS and CPVT as an adjuvant treatment to medication.
Methods: Between 2011-2018, 30 CSD (28 left, 2 right) were performed in 28 patients (21 LQTS and 7 CPVT, 26 genetically confirmed), aged 8 days-21y. Mean follow-up was 29 months. All were pharmacologically treated prior to surgery, 3 already had ICD. Indications for CSD in CPVT patients were prior cardiac arrest (3/7) and exertion or emotion-induced sustained VT despite medication (4/7). In LQTS, indications included syncope (4/21), VT despite medication (7/21), and high-risk of malignant arrhythmias (10/21) due to medication intolerance or noncompliance, family history of multiple sudden deaths, or T-wave alternation with QRS>650ms.CSD was performed via video-thoracoscopy in all; concomitant right sympatholysis was performed in 4 cases due to severe bilateral palmar hyperhidrosis.
Results: There have been neither intraoperative complications nor deaths. 4 patients had transient palpebral ptosis with mild miosis. Following CSD, 23/28 patients have been continuously monitored via ICD or ILR. After CSD, the vast majority (24/28) have been asymptomatic; three patients experienced syncope and one received an ICD appropriate shock. 7 had VT (3/7 for pharmacologic noncompliance), of which, 3 required an ICD and four improved with medication. 2 other had sporadic

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

non-sustained VT. At f-up, 5 high-risk patients received ICDs in primary prevention due to personal decision, but none have received shocks so far. 1 LQT7 patient with asymptomatic high-density ventricular arrhythmias has not improved at all despite multiple drug combinations and CSD.
Conclusion: CSD is, in experienced hands, an effective and safe technique in children as an adjuvant treatment for severe arrhythmias due to channelopathies. Following CSD, 85% of patients remain asymptomatic. Medication non-compliance is a prominent cause of post-CSD VT, though an ICD is occasionally required despite good compliance and CSD. In our experience, long-term remote monitoring is key for effective follow-up.

O1-6
Out-of-Hospital Cardiac Arrest in Infants, Children, and Adolescents in the Kyushu Area in Japan
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Background: In addition to Utstein-style reporting templates, information in detail that relate to out-of-hospital cardiac arrest (OHCA) may help prevent OHCA in pediatric patients.
Purpose: To clarify the characteristics of OHCA and determine risk factors associated with OHCA in the young.
Methods: The Council of the Kyushu Medical Association obtained data of patients with OHCA aged <20 years between 2012 and 2016 using questionnaires from local fire departments and school officials in Kyushu, Japan. The questionnaire asked about OHCA cases using the Utstein form and information on underlying diseases, prior activities, location of OHCA, and cause/situation. Subjects were divided in four groups: 0, 1–5, 6–11, and 12–19 years. One-month survival with a favorable neurological outcome was defined as a cerebral performance category of 1 or 2.
Results: A total of 605 cases were obtained, with 190, 118, 79, and 218 cases, respectively, in 0, 1–5, 6–11, and 12–19 age groups. The prevalence of a favorable outcome was 5%, 18%, 27%, and 21% (P<0.001) and prevalent cause/situations were during sleeping (71%), during sleeping (31%), water accident (26%), and suicide (24%), respectively, in each group.
OHCAs at home showed a significantly lower prevalence with a favorable outcome (24/388, 6%) than outside home/school (31/152, 20%, P<0.001) or at school (41/65, 63%, P<0.001). Exercise-related OHCA (31/48, 48%) and subjects with cardiovascular disease (21/55, 38%) showed a relatively high prevalence with a favorable outcome among prior activities and underlying diseases, respectively.
Multiple regression analysis with favorable outcome as the dependent variable showed that the presence of bystander CPR (odds ratio, 51.4; 95% CI, 23.2–113.9, P<0.001) and occurrence at school (5.5, 2.1–14.8, P=0.001) were independently predictive for a favorable outcome, and occurrence at home (0.33, 0.14–0.77, P=0.01) was a risk factor for an unfavorable outcome.
Conclusion: Different age-targeted strategies are required to reduce the prevalence of pediatric OHCA. Clarifying the etiology of infant death during sleep is required. Survival with a favorable outcome has been increasing in the school setting. To decrease the prevalence of OHCA at home, parents and/or the general population need CPR training.

O2-1
The Role of Cystatin C in Pulmonary Hypertensive Children
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Background and Objective: Cystatin C has been shown to be an important indicator of left heart failure and cardiovascular mortality in adults, and it has been strongly correlated with right ventricular ejection fraction. However, there is not enough data in childhood age group.
The aim of this study was to investigate the serum cystatin C levels of pediatric patients with pulmonary hypertension and to evaluate its relation with echocardiographic findings and to find out whether it may be used as a biomarker in the diagnosis and prognosis of pulmonary hypertension.
Materials and Methods: Twenty-two patients with pulmonary hypertension (10 patients with primary pulmonary hypertension and 12 patients with Eisenmenger syndrome) were included in the patient group. 19 patients who admitted with different complaints but had normal physical examination electrocardiography and echocardiographic findings were included in control group. Serum Cystatin C and proBNP levels of the participants were studied. Right ventricular ejection fraction, right ventricular end-diastolic volume, right ventricular end-systolic volume, right ventricular TEI index, TAPSE, and left ventricular eccentricity were measured by transthoracic echocardiography.
Results: The mean age of the patient and control group was 14.7, 15 years respectively. Cystatin C level was significantly higher in patients with pulmonary hypertension compared to the control group. When the patient group was sub-grouped as idiopathic pulmonary hypertension and Eisenmenger syndrome; Cystatin C level was significantly higher in patients with idiopathic pulmonary hypertension than Eisenmenger syndrome and control group.
In patients with pulmonary hypertension, right ventricular ejection fraction, TAPSE and left ventricular eccentricity measurements were found to be statistically significantly lower than control group. A statistically significant positive correlation was found between cystatin C and right ventricular TEI index.
Conclusion: Cystatin C levels are not affected by muscle mass, age and sex, they may be a superior biomarker than proBNP in pulmonary hypertension. Cystatin C levels were high and GFR was normal in patients with pulmonary hypertension. Despite our patient group is small, Cystatin C accurately correlates with RV pressure, function and morphology. Therefore, cystatin C may represent a novel biomarker in pulmonary hypertensive children.

O2-2
Ventricular-ventricular interaction in children with pulmonary hypertension: echocardiographic, hemodynamic, and disease severity (functional class) variables
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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Basics: Determination of ventricular-ventricular interaction (VVI) is an essential part of the echocardiographic examination in adults with pulmonary hypertension (PH), however, data from according pediatric studies is rare. We hypothesized that VVI variables in combination with left and right heart dimensions/function variables might indicate disease severity and progression in children with PH.
Methods: VVI variables as well as left heart, and right heart parameters were echocardiographically determined and correlated with NYHA functional class (FC), with N-terminal-pro brain natriuretic peptide (NT-proBNP), and invasive hemodynamic variables [i.e. pulmonary vascular resistance index (PVRi), ratio of systolic pulmonary arterial pressure divided by systolic pulmonary arterial pressure (sPAP/sSAP ratio)] in 57 children with PH (1-17 years; 7-84 kg, 24 female).
Results: The ratio of sPAP/sSAP and the PVRi correlated well with the left ventricular eccentricity index (LVEI), a marker of VVI, in our PH children ($p<0.001$). With increasing sPAP/sSAP ratio and increasing PVRi the left ventricular ejection fraction (LVEF) of our PH children significantly decreased ($p=0.001$). Patients with higher NYHA FC had lower LVEF and higher LVEI values ($p<0.001$). The sPAP/sSAP ratio and PVRi paralleled the increasing right ventricular (RV)/LV and right atrial (RA)/left atrial (LA) dimension ratios ($p<0.01$) of our PH patients. When stratified by NYHA-FC, with more severe exercise intolerance, RV and right atrial (RA) dimensions increased, and the systolic RV function variable tricuspid annular plane systolic excursion (TAPSE) decreased in our patients. Furthermore, the NT-proBNP values positively correlated with both, sPAP/sSAP ratio and NYHA FC ($p<0.01$).
Conclusions: The VVI variables LVEI and RV/LV dimension ratio were associated with clinical worsening, detrimental hemodynamics and increased NT-proBNP levels of our patients, thus emphasizing the importance of VVI in clinical management of pediatric PH.

O2-3
Prognostic value of serial measurements of NT-proBNP serum levels throughout the disease course in pediatric pulmonary arterial hypertension

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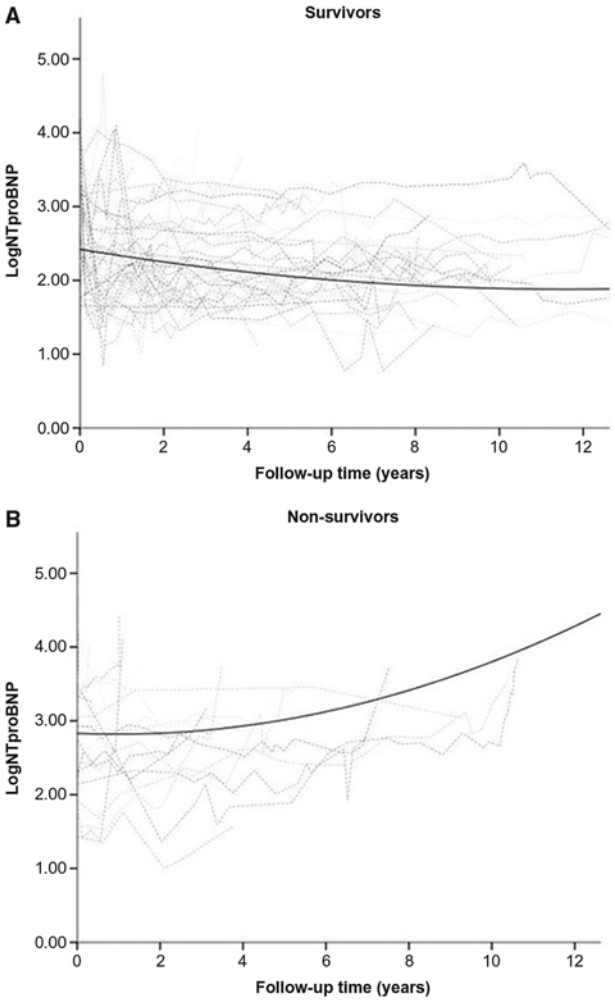
Introduction: N-terminal prohormone of brain natriuretic peptide (NT-proBNP), measured at time of diagnosis, has shown to be prognostic in children with pulmonary arterial hypertension (PAH) and treatment interventions are associated with changes in NT-proBNP levels. However, the prognostic value of serially measured NT-proBNP values has not been studied previously in PAH. We therefore aimed to determine whether serial measurements of NT-proBNP levels in children with PAH during the course of the disease have prognostic value for outcome and whether NT-proBNP levels throughout the disease course differ between survivors and non-survivors.
Methods: We included 82 pediatric PAH patients from the Dutch National Network for Pediatric Pulmonary Hypertension who had at least one follow-up measurement of NT-proBNP. The associations between serial measurements and concomitant variables known to represent disease severity were analyzed: World Health Organization-Functional Class (WHO-FC), 6-minute walking distance (6MWD), and tricuspid plane systolic excursion (TAPSE). Also, the courses of NT-proBNP levels were compared between survivors and non-survivors Additionally, we determined optimal cut-off values of NT-proBNP to predict risk for adverse outcome during follow up. Outcome was defined as (heart-)lung transplantation-free survival.
Results: Higher NT-proBNP levels correlated with higher WHO-FC, lower 6MWD and lower TAPSE at any time during the disease course ($p<0.001$ for all analyses). Each unit increase in NT-proBNP (log_{10}value) was associated with a 15.9 times higher risk for adverse outcome ($p<0.001$). A ten-fold increase in NT-proBNP level since baseline was associated with a 4.6 times increased risk of adverse outcome ($p<0.001$). Results remained significant after adjustment for age and sex. Non-survivors showed an exponential increase in NT-proBNP level over time in contrast to survivors who showed stable NT-pro-BNP levels ($p=0.005$) (see figure A and B).. A cut-off value for NT-proBNP of 700 ng/l and 1400 ng/l showed best discrimination for risk for an event in the long-term and short-term respectively.
Conclusions: Higher serum NT-proBNP values throughout the disease course are associated with worse disease severity and adverse outcome in pediatric PAH. These results suggest that monitoring NT-proBNP levels supports decision making regarding treatment strategies in pediatric PAH.



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

O2-4

Protein Losing Enteropathy after Fontan surgery - the Swedish experience

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Background: Protein-Losing Enteropathy (PLE) after operation with Total Cavo Pulmonary Connection (TCPC) has a prevalence between 5-15%. 5-year survival rate varies between 50-88%. PLE is characterized by enteric loss of proteins due to poorly understood mechanisms and treatment is difficult. This study presents prevalence, treatment and outcome of PLE in the Swedish cohort of TCPC patients.
Materials and methods: All patients with univentricular heart born in Sweden after Jan 1st, 1993 and operated with TCPC before 2017 (n=482) were reviewed and patients with PLE diagnosis before the age of 18 were included.
Results: Twenty-three patients with PLE were identified, corresponding to a prevalence of 4.8% with a median follow-up of 4.3 years (range 11 months – 15.2 years). In all cases PLE was suspected clinically and confirmed with elevated F-α1-antitrypsin. The most common morphological diagnoses were Hypoplastic Left Heart Syndrome (8/23) and tricuspid atresia (3/23). Eight patients had various forms of complex malformations. All patients had been operated in the neonatal period. The most common procedures were the Norwood procedure (9/23), pulmonary banding (6/23) and systemic-to-pulmonary shunts (6/23). Bidirectional Glenn, in some patients combined with other procedures was performed at median 6 months of age (range 3-19). The median age at TCPC was 2.7 years (range 1.2 – 6 years). Four patients had a lateral tunnel and 19 an extracardiac. Five patients had additional surgery concomitant with TCPC. The median interval between TCPC and PLE diagnosis was 11 months (range 3 months – 13.5 years). Pharmacological treatment was used in 19 pts and consisted of combinations of pulmonary vasodilators, heparin and budesonide. Eleven pts underwent 12 catheter interventions. Six patients underwent heart transplant. Nineteen patients are alive, 10 with continued pharmacological treatment, five in partial or complete remission and four after a heart transplant. Four pts have died corresponding to a mortality of 17%.
Conclusion: In this national study survival after the diagnosis of PLE was better compared to other studies. No single specific treatment was identified as successful, but transplantation was fairly common (26%). The study also indicates that remission of PLE occurs.

O2-5

Virus detection within endomyocardial biopsy in pediatric myocarditis: results from the German multi-center registry "MYKKE"

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Objectives: Endomyocardial biopsy (EMB) in pediatric patients is divergently discussed, but it might identify the underlying etiology of acute severe heart failure, trigger therapeutic interventions and might have a prognostic value. The aim of this prospective study was to analyze the histological pattern and viral distribution in EMB from pediatric patients with suspected myocarditis.
Methods: EMB derived from patients enrolled between September 2013 and December 2018 in the German prospective multicenter registry "MYKKE" were analyzed. EMB were studied with the use of histological (Dallas), immunohistological and molecularpathological analyzes; virus genome detection was done by polymerase chain reaction in the same laboratory.
Results: In 56% (205/364) of patients an EMB was performed; 11 patients had two EMB. All in all, 226 EMB reports were available for analysis. Median age at EMB was 12.9 (1.2-16.2) years. Biopsies derived from right ventricle in 84%, from left ventricle in 11% and both in 5%. In 72% (162/226) of the biopsies inflammatory reaction and therefore a myocarditis was positively detected: acute myocarditis (20%), subacute/chronic myocarditis (39%), status post myocarditis (12%), borderline myocarditis (4%), and hypersensitivity myocarditis (1%). In 5% no relevant inflammation and in 6% a DCM, 2% LVNC and 2% HCM was diagnosed; 8% had other diagnosis. In 44% (100/226) cardiotropic viruses in the myocardium could be detected (59% PBV19; 17% HHV6; 12% PVB19/HHV6; 4% Enterovirus; 3% CMV; 2% EBV, 1% HHV6/7, 1% HHV7). In 9% (21/226) simultaneously detection of virus within the myocardium and blood was ascertained, mostly in acute myocarditis (62%).
Conclusion: The rate of myocarditis (72%) and myocardial viral DNA (44%) detection in pediatric EMB for suspected myocarditis is unexpected high, while simultaneous virus detection in the blood could mostly not be proven. It seems that at a certain stage of symptomatic myocardial inflammation the virus titers within the blood have already been diminished, pointing towards timing as a

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

critical role for viral detection and the necessity of EMB as a diagnostic tool for potential antiviral or anti-inflammatory therapeutic interventions.

O2-6
Implementation of ABO-incompatible heart transplantation in Spain

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Introduction: Heart transplantation waiting list mortality for children under 1 year old in Spain is high (30%), despite a high national donation rate, due to the lack of donors for this age group. Average waiting time for children under 1y is 70 days and only around 4 heart transplants are performed per year.
Goal: Implementation of a national ABO-incompatible heart transplantation (ABOi-HT) program may decrease waiting list mortality and contribute to better outcomes.
Method: All 6 pediatric heart transplant centres in Spain agreed to start an ABOi-HT program in 2018. Pediatric recipient candidates for ABOi-HT should have isohemagglutinin titers ≤ 1:8. A policy of isohemagglutinin-free blood products administration for all potential recipients was implemented from listing time. Intraoperative exchange transfusion is performed if titers reach levels above 1:8. Immunosuppression was similar to ABO compatible HT. Allocation policies in the country for children with weight ≤ 15 kg were modified to give priority to highest risk patients without considering blood group compatibility.
Results: 11 patients have been included in the waiting list for ABOi-HT in a 1 y period, with a mean age of 5 months and a mean weight of 5,1 kg. Indications for HT were congenital heart disease (7, mainly LVHS in 6 cases),dilated cardiomyopathy (2) and other (2 RCM). 10 were transplanted, 4 patients underwent ABOi-HT and 6 ABO-compatible. Mean waiting list time was 44,8 days. Only 1 patient died while on the waiting list because of a neurological complication (mortality in the waiting list 9%). Two patients died after transplantation (1 of them ABOi-HT due to an widespread Aspergillus infection). The first three patients who received an ABOi-HT maintain low isohemagglutinins titers and good biventricular function after 11 months of follow-up.
Conclusion: ABOi -HT in Spain, with a policy of giving priority to highest urgency code patients regardless of blood group compatibility, resulted in a reduction of waiting list mortality, time on list and an increased probability of transplantation.

O3-1
Aortic arch restenosis rate after the Norwood procedure with polytetrafluorethylene patch

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Objectives: Children with hypoplastic left heart syndrome need a complex aortic arch reconstruction during the Norwood procedure using the native hypoplastic vessel, the pulmonary artery and an additional enlarging patch. Depending on the technique used, this area is prone to aortic arch restenosis which can lead to either catheter intervention or aortic arch reoperation.
Methods: Our retrospective single center study includes 104 patients who underwent a Norwood procedure using a curved polytetrafluorethylene patch cut out of a vascular prosthesis. The resulting patch is curved in two planes and enables an anatomically formed reconstruction. This patch was sewn into the inner curve of the neoaortic arch using 6/0 prolene. This material was earliest used in 4/2007 and similarly implanted as a homograft patch. The median age at operation was 8 days, median patient weight was 3,4 kg. 29% of the patients had an aortic atresia. Patients were followed until 8/2018 and the restenosis rate during follow up was assessed. Postoperative anatomy was examined echocardiographically and invasively before the bidirectional Glenn operation and Fontan procedure.
Results: The median bypass time was 201 min, median aortic cross clamp time was 71 min. Rethoracotomy because of early postoperative bleeding was necessary in 4% of the patients. The 30 days mortality rate was 4,8% and the late mortality rate during the follow up period of up to 11 years was 13,5%. During the median observation period of 5,2 years only one patient (1%) showed an early aortic arch restenosis that was treated with a balloon dilatation at the age of 12 months and later with a stent implantation. At the age of now 10 years she has no residual gradient. One patient underwent a correction of a kinking stenosis at the level of the ascending neoaorta at time of the Fontan procedure. Apart from this, there was no aortic reoperation in the patient population. Three patients are lost to follow up.
Conclusions: The curved polytetrafluorethylene patch in the neonatal aortic arch of a Norwood reconstruction shows an excellent longterm performance with a minimal reintervention rate during longterm follow up.

O3-2
Pulmonary artery sling repair: Single center experience with analysis of risk factors

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Objectives: Left pulmonary artery sling is a rare vascular anomaly causing respiratory failure. It is often associated with tracheal stenosis and intracardiac anomalies which mainly determine outcomes. There is a paucity of risk stratification in groups with pulmonary artery sling and similarly, there is very little correlation with coexistent hypoplasia of lung. The aim of this study is to review our experience and stratify risk factors for mortality and surgical outcome of all LPA sling repairs, including complex cases.
Methods: A monocentric retrospective analysis of patients between 2000 and 2017 was performed. Demographics, operative and perioperative data were collected. Univariate and multivariate analysis was performed for mortality and surgical outcome.
Results: Seventy nine consecutive children were operated. Median age at surgery was 5 months (interquartile range (IQR): 3-9.1 months). Surgical approaches include thoracotomy and sternotomy with a large amount of tracheal (n = 35) and intracardiac interventions (n=3), or a combination of both (n=28).
There were 7 early (8.8%) deaths. Two patients needed surgical revision of the left pulmonary artery anastomosis. The median intensive care stay and hospital stay were 11 (IQR: 9.2-24.8) and 17.9 (IQR: 4.3-19.8) days, and were considerably longer for those with associated tracheal surgery (p = 0.002). There were

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

three late deaths (3.8%) after 2, 10 and 17 months after surgery. Univariate analysis showed abnormal lung morphology and coexistent structural heart disease as statistically significant risk factors for mortality. Multivariate analysis revealed CPB time as an independent risk factor for overall mortality
Conclusion: Complex pulmonary artery sling repair can be performed with an acceptable surgical outcome. Abnormal lung morphology, associated cardiac lesions and long CPB time are risk factors for mortality. The approach should be patent tailored. Acceptable results are possible with simultaneous repair of the different lesions.

O3-3
Timing of the Arterial Switch Operation in Late Presenting D-Transposition Following Ductal Stenting
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Aims: Late presentation of (prenatally undiagnosed) transposition of the great arteries (D-TGA) is common in the developing world. Methods to retrain the left ventricle (LV) include two stage BT shunt with or without PA banding, and ductus arteriosus (DA) stenting. The timing of arterial switch surgery (ASO) after DA stenting is not certain. This study attempts to ascertain the duration required to retrain the (LV) using echocardiographic parameters.
Methods: D-TGA patients with regressed LV who underwent DA stenting were included. Echocardiographic parameters: indexed LV mass, LVPWd, LVEDV and sphericity index were measured serially on day 0 (day of DA stenting), 3, 5, 7 and weekly till surgery. Outcomes and complications were recorded.
Results: 12 patients underwent DA stenting (8M). Their median age and weight were 6.5 months (range 1 - 16 months) and 5.5 kg (2.4 - 9.2 kg) respectively. ASO was performed after a median period of 34 days (range 14 - 65 days). Serial echocardiographic parameters showed improvement in sphericity index, increased LV mass > 35 g/m^2 and increased LVPWd, allowing determination of optimal timing of ASO. 10 patients are doing well. One infant died of pneumonia 1 month post ASO, and another child died suddenly 3 years post ASO.
Conclusion: Transcatheter DA stenting is a good technique for retraining of regressed LV in late presenting D-TGA. Transthoracic echocardiographic parameters are useful in determining the optimal time of surgery

O3-4
Intramural coronary pattern in patients with transposition of the great arteries after an arterial switch operation
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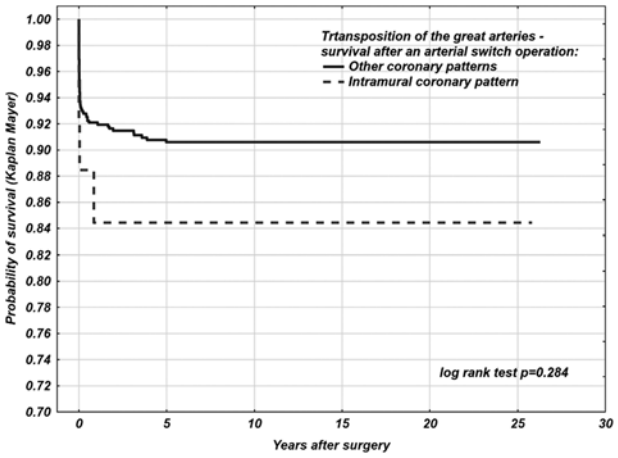
Introduction: Coronary complications are still the main reason for early mortality after an arterial switch operation(ASO). The high incidence of coronary anomalies in patients with transposition of the great arteries(TGA) may increase the difficulty of coronary transfer and among them intramural pattern was proved to be independent risk factor for early mortality. However recently published studies presented opposite results with no impact of this rare coronary variant on the survival rate.



The aim of this study was to assess the frequency of intramural coronary pattern in patients with TGA and its impact on early and late mortality after ASO. Additionally we presented all coronary arrangements associated with intramural pattern, which occurred in our cohort and the surgical technique to manage them successfully.
Patients and Methods: Since 1991 till 2017 we have performed 777 arterial switch operations with overall early mortality 6.9%, late mortality 2.5% and average follow up 12.65 years. This analyzed cohort include the initial learning curve with the higher mortality rate. All patients have detailed graphic and descriptive description of coronary anatomy in surgical report. They were reviewed, to describe the frequency of intramural coronary pattern, its impact on survival rate and the surgical technique used.
Results: Among 777 patients 262 had coronary anomalies (33.7%) and intramural pattern occurred in 26 patients (3.3%). This rare variant was associated in our cohort with 6 different coronary setups but most frequent was 1:0;2:LCA,RCA (10/26) with left coronary artery arising from right coronary sinus close to the posterior commissure and its intramural pattern between great vessels to the left atrioventricular groove. Combined early and late mortality were higher in patients with intramural coronary artery pattern(15.4%) in relation to other coronary variants(9.1%). This difference was not statistically significant (p=0.284, log-rank test, Figure), but the number of patients with intramural pattern makes statistics underpowered to confirm significance of the observed difference in mortality.
Conclusions: Intramural pattern associated with TGA still remains a surgical challenge but team experience seems to modify this risk factor. To reliably confirm or reject the significance of observed impact of this rare coronary variant multicenter data analysis is required.

O3-5
Systemic steroid therapy modifies postoperative lung injury after congenital cardiac surgery in neonates
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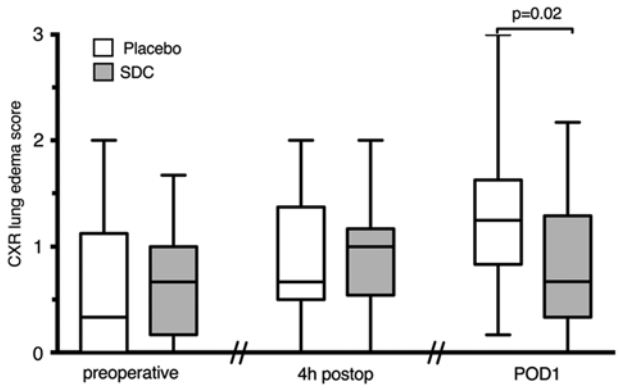
Introduction: Reperfusion after cardiopulmonary bypass (CPB) liberates inflammatory mediators and may cause lung injury with accumulation of excessive extravascular lung water. Systemic corticosteroid administration decreases inflammatory reaction. However, the data on whether systemic steroids attenuate postoperative inflammation and ischemia-reperfusion of the lungs after neonatal congenital cardiac surgery are scarce.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



Methods: We aimed to evaluate whether parenteral stress-dose corticosteroid (SDC) treatment reduces lung inflammation and ischemia-reperfusion injury after cardiac surgery and CPB. SDC and placebo were initiated perioperatively in 40 neonates in a double-blinded fashion. The SDC group received perioperative methylprednisolone at 2 mg/kg followed by hydrocortisone 0.2 mg/kg/h for 6-48 hours postoperatively, 0.1 mg/kg/h for the next 48 hours, and 0.05 mg/kg/h for the following 24 hours. The placebo group received saline in similar fashion. Postoperatively, lung edema in chest X-ray images, dynamic respiratory system compliance (Crs), oxygenation index, and PaO_2/FiO_2 -ratio were compared between the study groups.
Results: The CXR lung edema score was lower in the SDC group than in the placebo group on first postoperative day (POD1), but not preoperatively or 4 hours postoperatively (Figure 1). Furthermore, postoperative dynamic Crs was better in SDC group than in placebo group 4 – 6 hours postoperatively [7.3 (interquartile range (IQR) 6.2 – 8.9) vs 5.9 (IQR 5.0 – 6.7) ml/kPa/kg, p=0.006], on POD1 [7.3 (6.6 – 8.6) vs 5.3 (4.6 – 6.3), p=0.001], on POD2 [7.3 (6.5 – 8.4) vs 5.8 (5.0 – 6.5), p=0.001], and on POD3 [7.2 (6.6 – 8.8) vs 6.4 (5.4 – 6.6), p=0.005]. Postoperative oxygenation, however, showed no difference between SDC and placebo groups early postoperatively, or during POD1-POD3.
Conclusions: Intravenously administered corticosteroid reduced accumulation of radiographic lung edema and improved postoperative dynamic Crs. Thus, corticosteroids may attenuate postoperative lung injury after congenital cardiac surgery and CPB.

O3-6
Necrotizing enterocolitis in children with and without congenital heart defects

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Necrotizing enterocolitis (NEC) is a disease commonly found in preterm infants. Risk factors that contribute to the disease include asphyxia, apnea, hypotension, sepsis, and congenital heart diseases. Pneumoperitoneum is a well-accepted indication for surgery and in severe case peritoneal drainage.
Aim: To evaluate the survival of neonates with NEC and NEC with heart diseases (NEC+CHD) who underwent peritoneal drainage.

	N	DRAINAGE	SURVIVAL OF DRAINAGE	TOTAL	
NEC	109 (90%)	11 (10%)	1 (10%)	12	ref
NEC +CHD	131 (88%)	19 (12%)	7 (37%)	15	*p<0.01*
PDA	26 (80%)	7 (20%)	2 (29%)	33	
SHD	105 (90%)	12 (10%)	5 (42%)	117	

Patients and Methods: A retrospective study of 17 years (2000-17) involving 270 neonates with NEC (n=120) and NEC+CHD (n=150) subdivided in patent ductus arteriosus (PDA) and severe heart diseases. For the estimation of the relative risks (RR) and their 95% confidence intervals, simple log-binomial regression models (crude RR) and multiples (adjusted RR) were adjusted using weight, gestational age, Apgar 5th min, rupture of amniotic membrane and use of corticoids as covariates.
Results: There was a statistically significant difference between the crude RR [NEC / NEC+CHD] (95% CI) = 4.31 (2.86, 6.48) and adjusted RR [NEC / NEC+CHD] (95% CI) = 1.44 (1, 08, 1.94) (Fig); (RR 95%) = 0.54 (0.29; 0.99) and RR adjusted [severe] (95% CI) = 0.45 (0), and the presence of drainage (11/120 NEC and 19/150 NEC+CHD) crude RR [NEC / NEC+CHD] (95% CI) = 4.31 (2.86, 6.48) and adjusted RR [NEC / NEC+CHD] (95% CI) = 1.44 (1.08, 1.94).
Conclusion: NEC+CHD had greater number of survival when drained. Chronic hypoxemia of them before having pneumoperitoneum may explain this difference and may serve as a guideline for survival in indicating drainage.

O4-1
Preconception lifestyle intervention in obese women improves echocardiographic indices of cardiovascular function in their offspring: follow up of a randomised controlled trial

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Introduction: Maternal obesity has emerged as a risk factor for cardiovascular disease (CVD), as offspring of obese mothers are prone for hypertension, ischemic heart disease and type 2 diabetes. Animal studies and small observational human studies suggest that maternal obesity directly affects fetal cardiovascular development, which may explain the increased health risk in the offspring. This also suggest that a preconception lifestyle intervention among obese women may improve cardiovascular health in the offspring. In the present study we assess the effects of a preconception lifestyle intervention in obese women on echocardiographic indices of cardiovascular function in the offspring at the age of 6 years.
Methods: This study is embedded in the WOMB project (www.womb-project.nl), which is a follow up of a randomised controlled trial that included 577 obese sub/infertile women. A 6 months preconception lifestyle intervention aimed at weight loss prior to fertility care was given to the intervention group and a control group received fertility care as usual. We conducted complete transthoracic echocardiograms in the offspring at age 6-7 years using a Vivid E95 Ultrasound System (GE Healthcare, Australia). The clinician performing the echocardiograms and offline measurements was blinded

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

to group allocation. We used EchoPAC analysis software (GE Vingmed) for offline measurements of dimension, mass and stroke volume of the cardiac chambers.
Results: We included 44 children, mean age 6.1 years (SD 0.9), 57% girls. Children of women in the intervention group (n= 17) had a thinner interventricular septum (Z-score -0.6% [SD 0.7] vs 0.2% [SD 0.4], p <0.001) a lower left ventricular mass index (53.4 g/m2 [SD 9.1] vs 59.9 g/m2 [SD 7.0], p= 0.01) and an increased ejection fraction (60.9% [SD 3.5] vs. 56.6% [SD 4.6], p=0.004) compared to children of controls (n=27).
Conclusion: Preconception lifestyle intervention in obese women results in a thinner interventricular septum, a lower left ventricular mass and higher ejection fraction in the offspring at age 6, suggesting improved cardiac development and better cardiovascular function. This is the first direct human evidence of the effect of improving (pre)pregnancy maternal lifestyle to enhance cardiovascular function and potentially reduce CVD risk in the next generation.

O4-2
Serial assessment of somatic and cardiovascular growth in patients within the first decade after Fontan procedure

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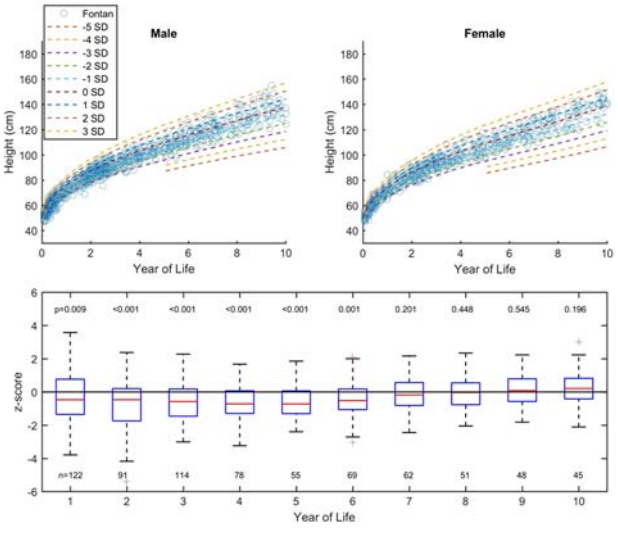
Background: Surgical modifications lead to improved long-term outcome after Fontan procedure. The effects of altered hemodynamic condition on somatic and cardiovascular growth in Fontan patients are still unclear. Aim of this study was to investigate the long-term growth regarding body weight, height as well as growth of both pulmonary arteries (LPA/RPA), single ventricle systemic valve annulus and single ventricle end-diastolic diameter (SVEDD) of Fontan patients with an extra cardiac conduit.
Methods: Anthropometric and echocardiographic data of 139 patients undergoing Fontan procedure from 1995 to 2018 at the University Children's Hospital Zurich were retrospectively analyzed. The first 10 years of somatic and cardiovascular growth were determined by z-scores. The time-point of normalization after Fontan procedure was assessed by a year-by-year comparison of z-scores using Wilcoxon signed rank test.



Results: The median age at Fontan procedure was 2.6 years (inter quartile range: 0.9 years) with a dominant left ventricle in 44% of all patients. In all patients a significantly lower weight and height was observed for the first 6 years of life (median z-score -0.4 to -0.9, p<0.05) compared with a healthy control population; afterwards, anthropometric parameters normalize. Similarly, RPA diameter is lower in the first 5 years of life (median z-score -1.1 to -0.6, p<0.05) and LPA in the first 8 years of life (median z-score -1.3 to -0.5, p<0.05). In contrast, SVEDD is increased in the first 4 years (median z-score 0.5 to 1.3, p<0.05) and normalizes afterwards. The median z-score of the systemic annulus is above 3 within the first 10 years of life and does not normalize. Somatic parameters are strongly correlated with cardiovascular ones over the study period (r >0.8, p<0.05).
Conclusions: Patients after Fontan procedure have a delayed somatic and cardiovascular growth (LPA/RPA), with a catch up in between the 5th to 8th year of life. The initially increased SVEDD normalizes after 4 years, the systemic annulus remains strongly enlarged over the first 10 years of life.

O4-3
Osteoprotegerin and RANKL serum concentrations in neonates of mothers with early-onset pre-eclampsia: comparison with preterm and term appropriate for gestational age neonates of normotensive mothers

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Introduction: A cytokine pathway of the tumour necrosis factor superfamily and its components Osteoprotegerin (OPG) and receptor activator of nuclear factor κB ligand RANKL have been implicated in the pathogenesis of cardiometabolic disease. Maternal pre-eclampsia is currently considered a risk factor for long-term cardiovascular complications in the mother and offspring. The OPG-RANKL axis function is altered in pregnant women with pre-eclampsia, however there is lack of data regarding OPG and RANKl concentratons in their neonates.
Methods: OPG and RANKL serum concentrations were measured in 28 premature neonates of mothers with pre-eclampsia, 28 appropriate for gestational age premature and 28 healthy term neonates of normotensive mothers.
Results: Neonates of mothers with early onset pre-eclampsia had significantly higher OPG levels compared to preterm and term neonates of normotensive mothers (Kruskall-Wallis p<0.0001). Also, RANKL concentrations of neonates of pre-eclamptic mothers exhibited significantly lower concentrations in comparison to preterm and term neonates of normotensive mothers (Kruskall-Wallis p=0.014). Linear regression analysis showed that pre-eclampsia (p<0.0001), birth weight SDS score (p=0.048) and antenatal steroid administration (p=0.034) were significant determinants of high OPG levels. Multivariable linear regression analysis showed that maternal pre-eclampsia was an independent predictor of increased diastolic and mean blood pressure in the offspring; however, its effect on systolic blood pressure was not significant
Conclusion: Early-onset pre-eclampsia, a relatively high birth weight and antenatal steroid administration affect OPG concentrations. Pre-eclampsia is an independent predictor of increased diastolic and mean blood pressure in the offspring and is possibly implicated in the 'fetal programming' of the cardiovascular system.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

O4-4
The influence of in utero exposure to metformin on body composition and cardiovascular phenotype in offspring; Metformin in Obese non diabetic Pregnant women (MOP) follow up

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Background/Basics: Maternal obesity and gestational weight gain have been associated with childhood obesity and long term adverse cardiometabolic outcome. In utero exposure to metformin in the MOP trial was associated with reduced gestational weight gain but had no influence on offspring birthweight. However, it remains unknown whether in utero metformin exposure can have long term influences on body composition and cardiovascular profile in the offspring.
Methods: Our study population consisted of 151 children from the MOP trial. Body composition was assessed by weight, body mass index (BMI), waist circumference, skinfold thickness and bioimpendance device (BIA-Biotekna) to estimate fat mass (FM), and free fat mass (FFM). Peripheral systolic (SBP) and diastolic blood pressure (DBP) were measured. Central aortic systolic (aSBP) and diastolic blood pressures (aDBP) were measured by Vicorder device using transfer function. Pulse pressure was calculated. Aortic pulse wave velocity between carotid to femoral segments was also assessed by Vicorder.
Results: The mean age of children was 3.9+/-1.0 years (76males, 75 females). From this cohort, 77 children were exposed to metformin. All body composition measurements were comparable between groups apart from gluteal circumference which was lower in metformin exposed group (56.7+/-6.6 vs 58.9+/-5.6cm in metformin vs placebo group, $p<0.05$). SBP and DBP were comparable between groups. Peripheral pulse pressure was marginally reduced in metformin exposed group compared to placebo (35.9+/-5.01 vs 37.5+/-5.5mmHg, p: 0.07). Central aSBP and pulse pressure were lower in children who were exposed to metformin compared to placebo (94.1+/-6.9 vs 96.3+/-6.9mmHg, $p<0.05$) respectively. Aortic pulse pressure was also lower in metformin exposed children compared to placebo (33.6+/-4.9 vs 35.5+/-5.2 mmHg, $p<0.05$) respectively. Aortic pulse wave velocity was comparable between groups.
Conclusion: This data suggests that in utero exposure to metformin does not have a beneficial effect on body composition in childhood. Central haemodynamics (aortic SBP and pulse pressure) were lower in metformin exposed group. As increases in central blood pressure have been associated with adverse cardiovascular outcome, longitudinal follow up is needed to assess whether metformin associated changes in central haemodynamics can translate in long term cardiovascular benefit.

O4-5
Prevalence of different forms of arterial hypertension and left ventricular hypertrophy in children after coarctation repair

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Introduction: Arterial hypertension (AH) is a one of the main complications of coarctation of aorta (CoA) and may persist or develop even after successful CoA repair. Main risk factors of persistence or development of AH after CoA repair is persistent or de novo recoarctation with low blood pressure (BP) difference between the right arm and lower leg. The aim of the study was to evaluate BP status and prevalence of left ventricular hypertrophy (LVH) in a cohort of children after CoA repair with a right arm and lower leg BP difference below 20 mmHg.
Methods: AH was diagnosed according to ESH2016 guidelines and BP status was further defined according to ABPM classification. LVH was defined as left ventricular mass index above the 95th percentile for age and sex.
Results: 67 patients (64.4% male) with a mean age of 12.4± 3.5 years, 3.1± 4.4 years after CoA repair were included in the study. 26 of 67 patients (38.8%) were normotensive. Overall, 26 of 67 (38.8%) had AH including 20 patients already treated with antihypertensive drugs and an additional 6 newly diagnosed as hypertensive. 8 (11.9%) had ambulatory prehypertension, 14 had white coat AH (WCH), 8 were in ambulatory prehypertension range and in 10 patients (14.9%) masked hypertension (MH) was diagnosed. In 3 of the 20 (15%) patients already on antihypertensive treatment BP was still in hypertensive range.
LVH was diagnosed in 25 of 67 (37.3%) patients including 3 of 9 (33.3%) hypertensive patients, 5 of 10 patients (50%) with MH, 8 normotensive, 3 with ambulatory prehypertension, and 6 patients with WCH (43%).
Conclusions: In children after correction of CoA but with low BP gradient between the right arm and lower leg, the prevalence of AH is high and affects 38.8% of patients. In addition, 14.9% of patients had MH. The prevalence of LVH was 37.3% in the group as a whole, including 50% of the patients with MH, and 43% of those with WCH. Our results indicate that children after CoA repair with persistent aortic blood flow abnormalities have high prevalence of AH and of LVH irrespective of BP phenotype.

O4-6
Health behaviors and cardiovascular disease risk factors in vegetarian adolescents

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Introduction: The prevalence of childhood obesity and metabolic syndrome is increasing worldwide, resulting in increased cardiovascular disease (CVD) risk. Achievement of optimal lifestyle behaviors is recommended. Previous studies in adults have suggested that vegetarians have lower adiposity, with less evidence in children. Therefore, we sought to determine the behaviors and CVD risk factors associated with self-identification as a vegetarian in adolescents.
Methods: Heart Niagara, Inc., Heart Healthy Schools Program has provided school-based universal risk factor screening for all grade 9 students in a defined geographic region since 1987 as a health curriculum enrichment program. The program includes assessment of family history, lifestyle behaviors, and CVD risk factors

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

based on questionnaires, anthropometric measurements, blood pressure and point-of-care lipid testing.
Results: From cross-sectional cohorts from 2009-2018, n=23,174 students (50.4% males) were included. There were 21,345 non-vegetarians (92%), 1,378 flexitarians (6%, eating mostly vegetables, but sometimes meat), and 451 vegetarians (2%). There were more flexitarians and vegetarians in girls (919 flexitarians (8%) and 336 vegetarians (3%) in girls; 459 flexitarians (4%) and 115 vegetarian (1%) in boys, $p < 0.001$). There was no significant differences between groups regarding BMI, waist to height ratio, and systolic blood pressure Z score, while flexitarians had slightly smaller height Z scores than non-vegetarians (-0.093, SE 0.028, $p = 0.02$). There was no difference in non-HDL cholesterol levels between groups, but HDL was lower in vegetarians (-0.042 mmol/L, SE 0.018). Flexitarians and vegetarians were more likely to 'skip meals' (OR 1.52, 95% CI 1.36–1.70, and OR 1.50, 95% CI 1.24–1.81, respectively). Flexitarians and vegetarians spent less time in moderate to vigorous exercise (-0.13 days/week, SE 0.056, and -0.27 days/week, SE 0.094, respectively). More flexitarians were smokers at examination (OR 1.81, 95% CI 1.22–2.68).
Conclusions: Vegetarianism is rarely reported by adolescents. It was not associated with reduced adiposity or cardiovascular risk factors, and in fact vegetarians had lower HDL and were more likely to report suboptimal dietary habits and less physical activity. These data suggest that vegetarianism may be paradoxically associated with increased CVD risk and worse lifestyle behaviors in adolescents.

O5-1
Novel LVNC genetic aetiology discovered by an original whole-exome sequencing data combinatory filtering method

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Background: Left ventricular non-compaction (LVNC) is a cardiomyopathy characterized by deep intra-trabecular recesses and prominent left ventricular trabeculae. This cardiomyopathy has remarkable phenotypic and genetic heterogeneity. Its molecular diagnosis via next generation sequencing is challenging due to the large amount of potential aetiological variants, especially in complex cases originating from mutations in various genes.
Objectives: This study sought to determine a novel diagnostic approach for identifying the molecular cause of LVNC in patients showing intra-familial phenotype variability and evoking a digenic aetiology.
Methods: We performed whole-exome sequencing (WES) in a three-generation family of patients affected by different degrees of LVNC. An enriched subset of 94 LVNC candidate genes was analysed in silico using an original WES data combinatory filtering method.
Results: We identified two novel heterozygous mutations, one in ACTC1 (c.740G>A; p.Gly247Asp) and another in ITGA7 (c.3280C>T; p.Gln1094Ter) which perfectly segregated with the phenotype. These variants underlined the disease's digenic origin, explaining intra-familial phenotype variability. We unambiguously determined that ITGA7 mutations lead to LVNC.
Conclusions: The results argues in favour of the final common hypothesis proposed as being involved in complex heterogeneous cardiovascular diseases. The genomic/computational approach presented here is an advantageous and efficient method for dissecting the molecular basis of LVNC's origin and we strongly recommend it for diagnostic/prognostic purposes. We have described the direct association of an ITGA7 mutation and LVNC pathophysiology for the first time, thereby enriching the repertoire of genes to be systematically analysed in LVNC patients.

O5-2
Yield of clinical screening for hypertrophic cardiomyopathy in child first-degree relatives: evidence for a change in paradigm

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Background: Hypertrophic Cardiomyopathy (HCM) is a heritable myocardial disease with age related penetrance. Current guidelines recommend clinical screening of relatives from the age of 10 years onwards but the clinical value of this approach has not been systematically evaluated
Methods and results: Anonymized, clinical data were collected from children referred for family screening between 1994-2017 following diagnosis of HCM in a first-degree relative. 1198 consecutive children (aged ≤ 18 years) from 594 families underwent serial evaluation [median 3.5 years (IQR, 1.2-7)]; 32 individuals met diagnostic criteria at baseline (median maximal LV wall thickness (MLVWT) 13mm (IQR, 8-21mm)) and 25 additional patients developed HCM over 4.6 years (IQR 2.8-7.1 years). Median age at first diagnosis was 10 years (IQR 4-13) and 41 (72%) were 12 years or younger at diagnosis (*figure*). Median age of affected

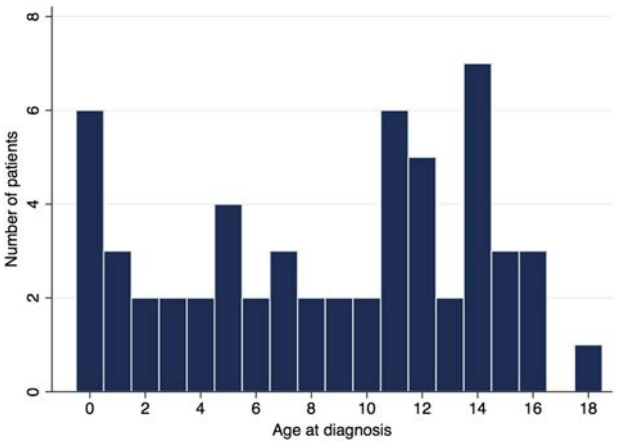

Figure.
Age of patients at diagnosis [≤12 years (72% n=41), ≤10 years (52%, n=30)]

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

patients at last follow up was 14 years (IQR 9.5-18.2). A family history of childhood HCM was more common in those patients diagnosed with HCM (n=32, 56%, VS n=257, 23% P <0.001). 18 patients (32%) were started on medication for symptoms, 2 (4%) underwent a septal myectomy, 14 (25%) received an implantable cardioverter defibrillator, 1 underwent cardiac transplantation, 2 had a resuscitated cardiac arrest and 1 died secondary to a cerebrovascular accident.
Conclusions: Almost 5% of first-degree child relatives from 8% of families undergoing screening meet diagnostic criteria for HCM at first or subsequent evaluations, with the majority presenting as pre-adolescents. The phenotype of familial HCM in childhood is varied and includes severe disease, suggesting that clinical screening should commence at a younger age

O5-3
Characteristics of in paediatric patients with hypertrophic cardiomyopathy -Effect of school-based cardiovascular screening on the prognosis

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Background: A school-based screening program to uncover cardiovascular disease is effective for early diagnosis and prevention of symptoms of inherited arrhythmia syndrome, particularly long QT syndrome, in Japan. However, little is known whether this program improves the prognosis of hypertrophic cardiomyopathy (HCM).
Purpose: To determine whether the screening program is effective for early diagnosis, prevention of symptoms or improvement of prognosis of childhood HCM.
Methods: A nation-wide study group for pediatric cardiomyopathy obtained data of patients aged <20 years who visited hospitals between 2000 and 2017. Data included age at diagnosis, sex, diagnostic event (by the screening program, or not), symptoms before and after diagnosis, echocardiographic data, presence or absence of medication or non-pharmacological treatment (surgical intervention including myectomy, implantable cardioverter defibrillator (ICD) implantation, etc), and poor prognosis. Poor prognosis was defined as the presence of death/coma, heart transplantation, and out-of-hospital cardiac arrest (OHCA).
Results: Among 375 patients with pediatric cardiomyopathy obtained, 134 had HCM. Of 96 and 38 patients with primary and secondary cases, respectively, 55 (57%) and seven (18%) cases were diagnosed with the screening program. Among the 96 primary cases, 10 patients had a poor prognosis; two patients died, one had transplantation, and nine had OHCA (overlapping was present). Fourteen patients received non-pharmacological treatment. Multivariate regression analysis using a poor prognosis as the dependent variable showed that the presence of symptoms before diagnosis (odds ratio, 9.88; 95% CI, 1.89–51.7; P<0.01) and a higher left ventricular wall thickness (interventricular septal + posterior wall thickness) (1.07, 1.01–1.14, P=0.03) were independent risk factors in primary HCM. No independent risk factors were found in secondary HCM. Screening with the program was not a predictive factor for lowering the prevalence of a poor prognosis.
Conclusion: Establishment of electrocardiographic and echocardiographic screening criteria for childhood HCM and early diagnosis and intervention before appearance of symptoms are essential for improving the prognosis of primary HCM in the pediatric population.

O5-4
Epigenetic Mechanisms Underlying Anthracycline-Induced Cardiotoxicity

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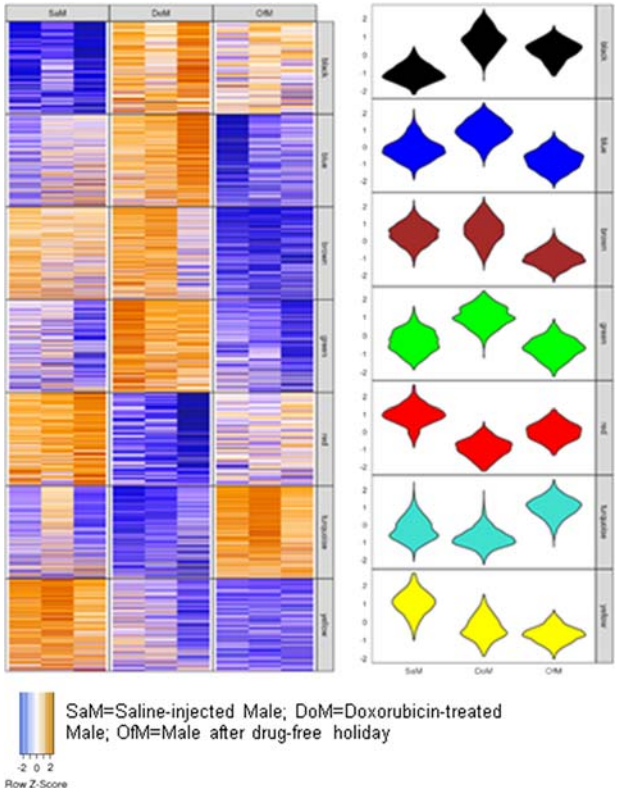
Introduction: Doxorubicin cardiotoxicity is consistently observed to develop chronically over time. We sought to develop a mouse model of pediatric anthracycline cardiotoxicity to test our hypothesis for an epigenetic mechanism (including epigenetic memory), which may be operating to explain delayed-onset anthracycline-induced cardiotoxicity.
Methods: Three week-old male and female C57Bl/6J mice were administered 5 weekly injections with doxorubicin (1mg/kg) (n=24) or saline (n=8). The dosage used was intended to closely mimic the clinical scenario, in place of the more frequently used protocols of single or multiple high dose anthracycline which induces acute toxicity in non-cardiac tissues and mortality. Half of the doxorubicin-treated (DOX) mice (n=12) and all controls (n=8) were sacrificed 1 week after the last injection; the rest of DOX mice (n=12) were given 4 weeks of drug-free holiday before



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

sacrifice. Echocardiographic measurements were obtained before the first injection, 1 week after the final injection, and before sacrifice. Gene expression profiles were assessed by RNA-sequencing, genome-wide DNA methylation by reduced representation bisulphite sequencing (RRBS), and chromatin accessibility by ATAC-seq. These analyses were performed on purified pools of isolated cardiac myocytes, procured using a published Langendorff-free method.
Results: With the selected dosing regimen, there was no death in either the DOX or control mice. Left ventricular chamber dimension and wall thickness as well as fractional shortening did not differ between DOX and controls, implying the lack of obvious functional insufficiency from this dose of DOX. Similarly, DOX mice did not differ in body weight, heart weights (HW), and HW-to-tibia length ratios compared with controls. Cardiomyocyte dimensions and myocardial interstitial fibrosis (using picrosirius red) were the same between groups. Despite the lack of echocardiographic and histological changes in DOX mice, RNA-sequencing analysis revealed profound and significant differentially expressed genes between the DOX mice and controls. Importantly, a significant proportion of downregulated and upregulated genes persisted, despite the period of drug-free holiday (Figure 1). Moreover, despite being taken off DOX, cardiomyocytes appeared to display new gene expression changes taking place even 4 weeks after stopping DOX. Detailed results of RNA-sequencing, RRBS and ATAC-Seq will be presented during the meeting.
Conclusion: These results suggest that cardiomyocytes display DOX-related epigenetic alterations, which may implicate the basis for delayed-onset anthracycline-induced cardiotoxicity.

O5-5
Prognosis and differences in mortality pattern between familial, sporadic and Noonan-syndrome associated hypertrophic cardiomyopathy presenting in childhood

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Introduction: Hypertrophic cardiomyopathy (HCM) presenting in childhood may be either familial, sporadic new mutations, or associated with the Noonan-group of syndromes. It has been reported that Noonan-associated HCM is a risk-factor for heart failure-related death but the size of risk has not been quantified compared with the other two types of childhood-HCM. Annual mortality is an in-correct measure to compare mortality in childhood HCM since mortality rate varies significantly in different age-ranges.
Methods: A Swedish national cohort of 144 patients diagnosed with HCM <19 yrs of age were categorized in familial non-syndrome associated HCM (Fam-HCM, n=67), HCM associated with Noonan-group of syndromes (NS-HCM, n=36), and sporadic new cases without syndrome-association with storage-disorders excluded (Spor-HCM, n=29). The remaining 12 could not be categorized with certainty and were not included in the analysis. Survival was compared with Kaplan-Meier survival analysis, and hazard of different types of cardiac mortality compared. There were 36 deaths: 24 sudden cardiac deaths (SCD), 9 heart-failure related, and 3 other cardiac deaths in the categorized groups.
Results: Overall long-term survival was not significantly different between Fam-HCM and NS-HCM (p=0.14), whereas Spor-HCM had significantly worse survival than Fam-HCM (p=0.026). The respective 10- and 20-yr survival were 88% and 84% for Fam-HCM, 80% and 70% for NS-HCM, and 70% and 56% for Spor-HCM. Mortality patterns were however very different with no heart-failure deaths in Fam-HCM, and 8/9 heart-failure deaths in NS-HCM. Hazard ratio for SCD was non-significantly higher in Spor-HCM than Fam-HCM (2.1 [95%CI 0.8-5.5], p=0.075).The hazard ratio of heart-failure death was 13.9 [3.1-61;p=0.0005)] comparing NS-HCM with Fam-HCM, and 4.0 [1.1-15;p=0.04] comparing NS-HCM with Spor-HCM. Genetic investigations in Spor-HCM revealed mutations in sarcomere genes in the majority. Right ventricular outflow-obstruction at rest occurred in 50% in NS-HCM, 10% Spor-HCM and 1.5% in Fam-HCM, whereas left ventricular outflow-obstruction at rest was present in 75%, 100% and 23% respectively.
Conclusions: NS-HCM is the dominant cause of heart-failure deaths in childhood HCM, whereas SCD occur in all three HCM-groups. Although Spor-HCM like Fam-HCM is commonly associated with sarcomere-protein mutations, the prognosis of de-novo mutations is worse than in patients with inherited mutations.

O5-6
The incidence of genetic abnormalities in fetuses with severe congenital heart defects

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Objectives: Overall survival of children with congenital heart defects (CHD) has improved significantly. Knowledge on additional morbidity, in particular genetic abnormalities, has become more important, as this is influences the prognosis of these cases. The aim of this study was to gather up-to-date information on the incidence of genetic abnormalities in fetuses with severe CHD to aid prenatal counselling.
Methods: The regional PRECOR database was used to identify all severe CHD cases (2012-2016). Aneuploidy cases were excluded from the study. Pre- and postnatal files and post-mortem reports were assessed to collect data on the presence of genetic abnormalities. The American College of Medical Genetics (ACMG)'s categories of clinical significance (pathogenic, likely pathogenic, uncertain significance, likely benign or benign) were used to interpret all genetic anomalies encountered. We assessed the incidence of genetic anomalies amongst all CHD subjects, specific types of CHD and the association with extra-cardiac malformations (ECM).
Results: Genetic abnormalities were encountered in 178/710 (25.1%) of subjects with severe CHD. In 27.5% genetic testing was not performed, as there was no clinical suspicion for a genetic syndrome. Pathogenic alterations, likely pathogenic alterations and variants of uncertain significance (VOUS) were observed in 13.8%, 1.8% and 5.6% of all CHD subjects respectively. Common genetic anomalies comprised 22q11 deletion (4.2%), CHARGE (0.7%), Kabuki (0.4%) and Noonan syndrome

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

(0.4%). Genetic abnormalities were most frequently encountered amongst common arterial trunk (37.5%), atrioventricular septal defect (35.7%) and Tetralogy of Fallot (27,6%) cases. The risk of genetic abnormalities was significantly lower for isolated cases (21.2%) compared to cases with additional ECMs (38.8%) ($p<0.001$).
Conclusions: The yield of genetic and syndromic abnormalities amongst fetuses with severe CHD and a normal karyotype was 25.1%, of which 15.6% comprised pathogenic or likely pathogenic anomalies. This information is valuable for prenatal counselling and postnatal care management in these subjects, especially in the presence of ECMs.

O6-1
Pediatric Normal Values for Pulsed Doppler Velocities, Times, and Velocity Time Integrals for Semilunar Valves and Great Vessels

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Background: Doppler velocities are commonly employed for functional evaluation of semilunar valves and great vessels, however, pediatric nomograms are limited. Our aim was to prospectively establish pediatric nomograms for Doppler velocities in the great vessels at different sites in a large pediatric cohort.
Methods: We prospectively studied 912 healthy Caucasian Italian children (age 31 days-17 years, 48.6% female). Echocardiographic measurements included: pulsed Doppler velocities (cm/s), acceleration/deceleration times (ms), ejection times (ms), and velocity time integrals (VTI; cm) at multiple sites including the aortic valve, pulmonary valve, and aortic arch. Sub-aortic, pulmonary annulus and aortic isthmus diameters were measured, and stroke volume (ml) and cardiac output (L/min) derived at all sites. Age, weight, height, heart rate (HR), and body surface area (BSA) were used as independent variables in different analyses to predict mean values of each measurement.

Table 1a: Aorta Coefficients for regression equations relating echocardiographic measurements and BSA, the Standard Error of the Estimate, the determination coefficient. Normality test: Shapiro-Wilk and Lilliefors (Kolmogorov-Smirnov). Heteroscedasticity test (White test and Breusch-Pagan test). BSA HAYCOCK. (ln[y] = a + b*ln[x]); Z value = (ln[Measurement] –(Intercept + B*ln[BSA]))/$\sqrt{}$MSE

Measurement	Intercept	B	SEE ($\sqrt{}$MSE)	R2	SW	KS	BP	W
Ao VTI (cm)	3.160	0.353	0.188	0.459	0.067	0.193	0.468	0.085
Ao ejection time (ms)	5.687	0.209	0.112	0.459	0.062	0.087	0.074	0.264
Ao Acc Time (AT) (ms)	4.309	0.314	0.282	0.233	<0.001	<0.001	<0.001	<0.001
Ao Dec Time (DT) (ms)	5.383	0.180	0.162	0.230	<0.001	<0.001	0.184	0.724
AO SV (ml)	3.686	1.324	0.243	0.878	0.868	0.200	0.064	0.053
AO CO (L/min)	1.224	0.902	0.247	0.759	0.601	0.200	0.971	0.234

Table 1b: Pulmonary artery:BSA HAYCOCK. (ln[y] = a + b*ln[x]); Z value = (ln[Measurement] –(Intercept + B*ln[BSA]))/$\sqrt{}$MSE

Measurement	Intercept	B	SEE ($\sqrt{}$MSE)	R2	SW	KS	BP	W
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AO CO (L/min)	1.224	0.902	0.247	0.759	0.601	0.200	0.971	0.234

Results: At all sample points the VTI, ejection time, diameter, SV and CO were positively correlated with BSA and age, but negatively correlated with HR ($r \geq 0.6$, $p<0.001$). Models with exponential (ln[y]=a+b*ln[x]), linear (y=a+b*x) or cubic ($y=a+b1*x+b2*x^2+b3*x_3$), equations showed the best results. For all parameters, the association with BSA was stronger than for HR or age, so BSA was used for normalization. Predicted values and Z-score boundaries by BSA are provided.
Conclusions: We report pediatric echocardiographic normative data for Doppler velocities, acceleration/deceleration times, and VTI for semilunar valves and great vessels in a large pediatric population.Our data would serve as a baseline Doppler evaluation in children with congenital heart disease.

O6-2
Left ventricular strain and strain rate during submaximal semisupine bicycle exercise stress echocardiography in healthy adolescents and young adults – systematic protocol and reference values

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Objectives: Combining stress echocardiography with strain analysis is a promising approach for early detection of subclinical cardiac dysfunction not apparent at rest. Study protocols differ considerably, data on normal myocardial strain and strain rate response to exercise in adolescents and young adults is contradictory and limited. The aim of this study was to propose a systematic standardized protocol for semisupine bicycle stress echocardiography and to provide corresponding reference values of left ventricular (LV) two dimensional speckle tracking echocardiography (2D STE) strain and strain rate (SR) in adolescents and young adults.
Methods: 50 healthy adolescents and young adults (mean age 17.8 ±3.2 years, 44% female) were prospectively assessed. Images were acquired at rest, low stress, submaximal stress and post exercise. Optimal image quality for offline strain analysis was pursued and image quality was rated. Global longitudinal, apical 4-/2-/3-chamber longitudinal and short axis circumferential strain and SR were analyzed using vendor-independent software. Interobserver variability was assessed.
Results: Strain and SR increased during progressive exercise stress. Mean LV global longitudinal strain was -20.4±1.3%, SR -1.1 ±0.15/s at rest (heart rate (HR) 79.4±12.0/min), increasing to -22.6±1.6% and -1.5±0.16/s at low stress level (HR 117.1±8.7/min) and -23.7±1.1% and -1.9±0.29/s at submaximal stress level (HR 154.2±7.0min) respectively, returning to -20.6±1.4% and -1.2±0.16/s post exercise (HR 90.1±9.4). Restriction on submaximal stress level ensured adequate image quality allowing 2D-STE strain analysis in >95% of loops recorded in apical 4-chamber and short axis view and in >80% for apical 2- and 3- chamber view. Interobserver variability was within acceptable limits and did not reveal significant interobserver bias.
Conclusions: This study provides a systematic, standardized protocol and corresponding reference data for 2D-LV STE derived strain and strain rate during semisupine bicycle stress testing in adolescents and young adults. According to our results, global longitudinal strain and strain rate might be the most comprehensible parameters for cross-sectional studies. This might help to interpret future stress strain studies for the detection of subclinical LV dysfunction.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

O6-3

Impact of Obesity on Left Ventricular Thickness in Children with Hypertrophic Cardiomyopathy.

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Introduction: Hypertrophic cardiomyopathy (HCM) is a genetic heart disease with diverse natural history. The hallmark of HCM is severe left ventricular hypertrophy (LVH). In adults with HCM, obesity additionally increases LVH. It is not known whether obesity can lead to further LVH in children with HCM.
Hypothesis: Obesity is associated with additional LVH in children with HCM.
Methods: Echocardiogaphic LV dimensions were determined from 2D and/or M-mode images according to established criteria in 504 children ≥2 and ≤20 years of age with phenotypic sarcomeric HCM. Children with HCM associated with syndromes and storage disorders were excluded. Genotype-positive, phenotype-negative patients were excluded. Echocardiographic measurements of interventricular septal thickness (IVST) and posterior wall thickness (PWT), and patients' weight and height at the time of the echo were collected. Obesity was defined as a body mass index (BMI) ≥95th percentile for age and sex, as defined by CDC guidelines. IVST data was available for 498 and PWT data for 484 patients.
Results: Patient age ranged from 2 to 20 years (mean ± SD, 12.5 ± 3.9) and 340 (68%) were males. Overall, patient BMI ranged from 7-50 (22.7 ± 6.1). Obesity (BMI 18-50, mean 29.1) was present in 140 children whose age ranged from 2-19.6 (11.3 ± 4.1). The overall mean IVST was 20.5 ± 9.6 mm and the overall mean PWT was 11.0 ± 8.4 mm. The mean IVST in the obese patients was 21.6 ± 10.0 mm and mean PWT was 13.3± 14.7 mm. The mean IVST in the non-obese patients was 20.1 ± 9.5 mm and mean PWT was 10.4 ± 4.3 mm. Obesity was not significantly associated with IVST (p=0.12), but was associated with increased PWT (0.0011). The average predicted value of PWT for a non-obese child was 10.4 mm but was 13.3 mm if obese.
Conclusions: Presence of obesity does appear to influence LV thickness in children with HCM. Obesity is associated with increased PWT. However, IVST appears to be independent of obesity. These findings could have implications in risk stratification strategies utilized to identify children at risk for sudden cardiac death. Whether obesity and its impact on LVH influences clinical outcomes in children with HCM needs to be confirmed by further studies.

O6-4

Increased Cancer Incidence Following 15 Years after Cardiac Catheterization in Infants under 1 Year between 1980 and 1998 in a Single Center

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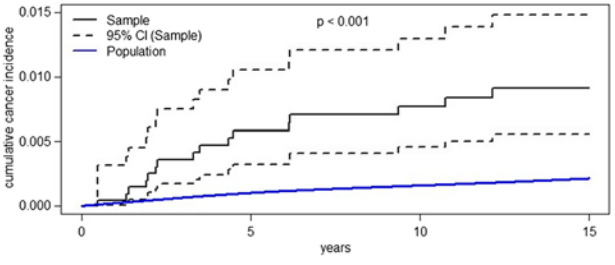
Objectives: Cardiac catheterization is associated with a significant radiation exposure in the pediatric patient population, bearing an excess risk of cancer development. In this study we aimed to assess the risk of cancer in a cohort of children who underwent cardiac catherization in our institution in the first year of life.
Methods: In our retrospective study we included 2770 infants, of whom 7.8% had trisomy 21, without known malignant disease, who underwent cardiac catheterization in our institution at the age of less than one year between the 1st of January 1980 and the 31st December 1998. In this cohort, newly diagnosed cancer, occurring in the first 15 years of life was detected through record linkage to the German Childhood Cancer Registry (GCCR). Cancer risk was assessed and compared to the GCCR incidence rates for the general German population of children less than 15 years calculating the standardized incidence ratio (SIR). Subgroup analysis was performed for patients with trisomy 21, comparing cancer incidence with data from the Danish Cytogenic Register for trisomy 21.



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Effective radiation doses were calculated by Monte Carlo (MC) simulations for each tumor patient and for 60 randomly selected patients out of the whole cohort who did not develop cancer. MC simulations were performed after adjustment for patients' weight and length on the basis of registered dose area products.
Results: A total of 24472.5 person years were analyzed. Sixteen patients developed malignant tumors until the age of 15 without predominance of a specific cancer type. The number of expected cancer cases until the age of 15 is 3.64 (SIR) = 4.4, 95%-CI: 2.5-7.2, p<0.001) (see Figure). The proportion of patients with trisomy 21 was higher among patients with cancer (4/16 = 25%) than in the studied cohort (7.8%, p = 0.034). All 4 patients had leukaemia or myelodysplastic syndrome.
Median effective radiation dose in 15 patients with cancer was higher (65.1 mSv, range: 0.8-242.3 mSv) than in controls, although statistically not significant (28.5 mSv, range: 0.0-750.0, p>0.1).
Conclusions: Cardiac catherization in the first year of life between 1980 and 1998 was associated with a significantly increased cancer risk.

O6-5
New Screening Tool for Aortic Root Dilatationin Children with Marfan Syndrome and Marfan-like Disorders

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Background: The primary role of pediatric cardiologist, consulting patients with a suspicion or diagnosis of a genetically determined connective tissue disease (such as Marfan, Ehlers-Danlos or Loeys-Dietz syndromes) is to assess whether aortic root is dilated. Patient's age, gender and body surface area influence aortic root diameter, therefore, it is not possible to establish its single normal range for entire population. Thus it is necessary to assess aortic root diameter with special nomograms and express it in z-score. Z-score calculations are time-consuming and could be troublesome, if used infrequently. This study was aimed at introducing a simple screening method for identification aortic root dilatation in children.
Methods: Study population consisted of 190 children (3 months -18 years) with the diagnosis of Marfan syndrome or Marfan-like disorders. Aortic Root ratio (ARr) was created, which was a simple quotient of aortic root diameter to patient's height (Fig.1). The value of ARr in each patient was confronted with the results obtained using three most widespread z-score calculators (based on nomograms by Gautier et al., Pettersen et al. and Cantinotti et al.). The ROC curves analysis was employed to evaluate the predictive value of ARr to identify aortic root dilatation and to determine the optimal cut-off value of ARr.

$$\frac{\text{aortic root diameter (mm)}}{\text{patient's height (cm)}} * 100$$

Figure 1.
Formula for calculation ARr.

Results: Surprisingly, we revealed that three commonly used z-score calculators were not perfectly concurring with each other – in as many as 5.8% of patients the results (dilated or non-dilated aortic root) were divergent between them. The calculated optimal cut-off value of ARr was ≥18.7. At that cut-off point the sensitivity of ARr ranged from 88.3% to 100% and the specificity ranged from 94% to 97.8% (depending on z-score calculator applied for comparisons). In all patients, in whom ARr at that cut-off point failed to identify aortic root dilatation, the results were divergent using different z-score calculators. At the cut-off point of ≥18.0 the sensitivity of 100% was achieved for all z-score calculators.
Conclusions: ARr allows for rapid and precise screening for aortic root dilatation in children. Unlike the classic analysis, ARr doesn't necessitate access to any nomograms or on-line calculators.

O6-6
Decreased Total Lung Capacity in Patients with Tetralogy of Fallot

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Objectives: Patients with repaired tetralogy of Fallot (TOF) had an inborn right heart obstructive congenital heart disease, leading to possible lung deficits such as impaired forced volumes (FVC and FEV1) or total lung capacity (TLC). This study aims to prove this hypothesis in children, adolescents or young adults with repaired TOF.
Methods: From April to November 2018 forty-one patients with TOF (16.2 ± 5.0 years, 39% girls) underwent a spirometry and bodyplethysmography. Results are expressed in relation to their reference values as %predicted (mean ± SD). Data were tested against 100% with a one sample T-Test.
Results: In TOF FVC %predicted TOFs was reduced to 84 ± 15%, FEV1%predicted 83 ± 14% and TLC %predicted 89 ± 11% (p<0.001 for all). There was no increase in RV %predicted (107 ± 35%, p=0.208) and FEV1/FVC %predicted (100 ± 8%, p=0.988)
Conclusions: FVC, FEV1 and TLC are reduced in TOF patients. These findings suggest that this CHD affects patients' lung development. Further data are needed to evaluate whether this is due to an inborn lung defect associated to TOF, a fetal delay in development or/and a result of the early chest operation.

O7-1
Causes and outcomes of pulmonary hypertensions confirmed with right heart cath in infants younger than 6 months

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Background: The epidemiology of pulmonary hypertension in infants younger than 6 months is poorly known as they are either excluded of registries or do not have right heart catheterization to characterize their hemodynamic.
Objectives: We retrospectively reviewed all RHC performed over a period of 8 years in infants less than 6 months of age with the purpose to evaluate pulmonary hemodynamic. We define PH as mean PAP > 25 mmHg and PVRi > 3 WU.m2 for those with biventricular physiology with or without shunt. Any value of pulmonary wedge pressure was accepted. For segmental PH, only mPAP was

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

considered. Patients were classified according to the Nice classification.
Results: 210 infants < 6 months were included into the study. 184 had PH confirmed. Causes of PH and/or indications for RHC were: group 1 (Pulmonary arterial hypertension) - evaluation of left-to-right shunts for operability or pulmonary blood flow contribution to clinical condition in 69 (37.5%), idiopathic PAH in 3, PVOD in 1, associated PAH-other in 5 (total 42.4%); group 2 (Postcapillary PH) - diagnosis of pulmonary veins anomalies in 29 (15.7%), and PH in obstructive left heart diseases in 26 (14.1%); group 5 (Multifactorial) - scimitar syndrome in 10; PH after the ASO for TGA in 6; complex congenital heart diseases and segmental PH in 14 (total 16.3%); group 3 (developmental lung diseases) isolated in 5 and with coincidental CHD in 16 (total 11.4%). There was one RHC related death due to sepsis. One-year mortality in the total population at one year was 35/184 (19%) and mainly related to persisting PH. 76% of survivors had normal pulmonary pressure at one year of age either after surgical correction of CHD or improvement of their lung disease. 37 patients (24% of survivors) had PH at one year of age.
Conclusion: PH in young infants is mainly associated with congenital heart diseases (CHD) with all groups being represented. Idiopathic PAH and developmental lung disease are rare in this age group. The transitory aspect of PH is frequent in all PH-CHD groups. Persisting PH at one year is present in a fourth of the population.

O7-2
A novel atrial flow egulator (AFR) device to control and assure blood flow after balloon atrioseptostomy (BAS) for severe, endstage pulmonary arterial Hypertension (PA)

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Introduction: PAH may progress to its final, pre-lung-transplant stage despite high-dose, multiple combined medications. A BAS may be offered to those patients that need to wait, need to improve to become elegible, or are remain inelegible to offload the venous system and right ventricle, and provide much needed extra systemic blood flow. However, size of BAS and hence resulting shunt flow are difficult to control, and BAS tend to close, with needing repeat procedures, thus leading to some neglection of this otherwise hemodynamically useful and important Intervention in the recent years.
Methods: a purpose made device was developed (Occlutech) that uses the self-centering ability of ASD-closure devices but has a fenestration tunnel of different diameters (4-10 mm) to be implanted into the defect produced by preceeding BAS. A formal study protocol to achieve European CE-marking has been set up (see "PROPHET" trial at www.clinicaltrials.com), and recruitment is ongoing and europe-wide.
Patients: 8 adult patients (39-62 years) with multi-medicated terminal PAH were included and received the AFR device without complications in all. AFR remained in situ and patent in all, with 2 patients > 6 months of follow-up time. 4 patients dropped out < 6 months due to disease progression. One patient required exchange of AFR device for one with a smaller fenestration. The remaining patients all showed improvement as judged by organ parameters including NT-BNP, and clinical state, and 1 of these, the AFR device served as a bridge to transplantation with the patient receiving a lung transplant a month after intervention, with the patient remaining well to date and the - now left to right shunting- AFR device safely left in situ.
Conclusion: While the AFR-implantation after BAS is a swift and uncomplicated intervention, this is the first time that a known controlled interatrial right to left shunt flow can be achieved and appreciated in its effects within PAH treatment. Choice of fenestration width is of highest importance. Continuation of our PROPHET study is warranted to further establish the use of this important interventional hemodynamic treatment option in refractory final PAH.

O7-3
Early Pulmonary Hypertension in Extremely Premature born Infants: Results from a prospective cohort study (Neolifes-Heart)

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Introduction: Early pulmonary hypertension (PH) is known to complicate the course of extreme preterm born infants. However, prevalence of early PH in this population is insufficiently known. The aim of this prospective cohort study was to determine prevalence and risk factors of early PH. Also its association with subsequent development of bronchopulmonary dysplasia (BPD) was assessed.
Methods: In a single-centre prospective cohort study in the University Medical Centre Groningen from June 2015 until November 2018, infants were included with a gestational age<30 weeks and/or a birth weight<1000 grams. Echocardiographic assessment for the determination of PH was performed in the first week of life (day 3-10). Neonatal and maternal clinical data (e.g. gender, gestational age, birth weight, Apgar score, presence of patent ductus arteriosus (PDA), presence of comorbidities, preeclampsia, oligohydramnios, intra-uterine growth restriction, antenatal corticosteroids-use) were collected. BPD status and its severity were assessed at 36 weeks postmenstrual age (PMA).
Results: In total 94 infants were included. Early echocardiographic revealed PH in 40 of these infants (43%). Of the infants with PH, 26 (65%) infants subsequently developed BPD, where 11/26 (42%) infants developed severe BPD. In comparison, 20 of the 54 infants (37%) without early PH subsequently developed BPD, and 3/20 (15%) developed severe BPD. The presence of early PH was associated with the subsequent development of BPD (odds ratio (OR):5.01;1.89–14.68), and even stronger for severe BPD (OR:14.14;3.41–76.80). The presence of PH was associated with a low Apgar score after 1 minute, the presence of a PDA (OR:6.19;2.10–22.87) and the absence of antenatal maternal corticosteroid-use (OR:0.31;0.09–0.95). In total 8 infants died during the study, of which 5 infants had early PH.
Conclusion: Over 40% of infants born after a gestational age<30 weeks and/or a birth weight<1000 grams, presented with PH in the first 10 days of life. Low 1-min Apgar score, the presence of a PDA and the absence of antenatal corticosteroids, appeared risk factors for the presence of early PH. Furthermore, the presence of early PH was associated with the subsequent development of BPD, especially severe BPD at the age of 36 weeks PMA.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

O7-4

Von Willebrand factor reflects Fontan pathophysiology and strongly predicts the all-cause mortality

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Introduction and Objectives: Von Willebrand factor (vWF) has prognostic value not only in patients with heart failure but also in those with liver cirrhosis. Liver fibrosis as well as heart failure are major late complications that predict poor prognosis in patients late after Fontan operation. The purpose of the present study was to clarify clinical significance of measuring plasma levels of vWF antigen (vWF:Ag), including the prognostic value in Fontan patients.
Methods: We measured vWF:Ag (%) in consecutive 278 Fontan patients and compared the results with the clinical profiles, including hemodynamics and prognosis.
Results: Plasma vWF:Ag was 144±48 (normal range: 55-190%) and 37 patients (13%) showed high levels of vWF:Ag (≥190). Male gender, late Fontan operation, greater New York Heart Association (NYHA) class, elevated plasma neurohormons (norepinephrine, brain natriuretic peptide: BNP, renin activity, and aldosterone: PAC), protein losing enteropathy, elevated central venous pressure, low systemic pressure, hypoxia, use of diuretics and anti-arrhythmias and low liver synthetic function (low plasma levels of low albumin and cholinesterase) were associated with high vWF:Ag ($p<0.05$-0.0001). Of these, low albumin, greater NYHA class and high PAC were independently associated with a high vWF:Ag ($p<0.05$-0.01). During the follow-up, 51 clinical events, including 9 deaths, occurred. High vWF:Ag predicted the clinical events ($p<0.01$), especially the all-cause mortality (hazard ratio: 1.3 per 10, $p<0.0001$), with being independent of significant prognostic value of BNP ($p<0.0001$). Patients with high vWF:Ag (≥218) and BNP (≥65 pg/ml) had a marked high hazard of 166 for all-cause mortality ($p<0.0001$).
Conclusions: Low albumin and high PAC were associated with a high vWF:Ag which strongly predicted all-cause mortality independent of hemodynamics, including BNP, in Fontan patients. Thus, vWF is a novel clinically useful biomarker of Fontan pathophysiology.

O7-5

Enalapril orodispersible minitablets (ODMTs) for children with heart failure – a successful EU-funded drug development program for children

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Introduction: Until today, European and American children with chronic heart failure do virtually not benefit from approved drugs despite the major successes of the US paediatric legislation initiative in 1997 and the European paediatric regulation in 2007. The EU-funded LENA consortium (FP7 Grant agreement no 602295) has bridged this gap by generating all data necessary for a paediatric-use marketing authorization (PUMA) application for the angiotensin-converting enzyme (ACE) inhibitor enalapril to treat paediatric heart failure patients.
Methods and Results: On the bases of good manufacturing, as well as good clinical and laboratory practices, investigator-driven clinical trials in 24 healthy volunteers, and in 102 children aged from birth to 12 years (72% below 1 year of age) demonstrated substantial bioavailability of the enalapril orodispersible minitablets (ODMT) as well as safe, simple and reliable use. The ODMTs were highly accepted by patients, parents and medical staff. The ODMTs remove the need for off-label enalapril use in extemporaneous formulations, including liquids, capsules or crushed tablets in children below the age of 6 years or 20 kg and age-appropriate safe dosages of enalapril are now established.
The LENA consortium has successfully developed a risk-adapted quality management system for an academic consortium. In addition, they developed an innovative active learning tool for particularly critical processes in these trials. This enabled the LENA team to fulfil all applicable ethical and regulatory standards for study performance and reliable data generation in their pharmacokinetic and pharmacodynamic investigations.
Conclusions: As ACE-inhibitors are regarded as first line treatment and enalapril was prioritized by the EMA to be developed for paediatric heart failure treatment, the LENA enalapril ODMTs provide a substantial step towards the safe and reliable treatment of children with heart failure.
The research leading to these results has received funding from the European Union Seventh Framework Programme (FP7/2007-2013) under grant agreement n°602295 (LENA).

O7-6

Conversion to everolimus - a treatment option in pediatric heart transplant recipients: impact on CAV, renal function and cardiovascular risk factors

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Introduction: Cardiac allograft vasculopathy (CAV) is considered to be a major cause impairing the long-term survival after heart transplant. Additional side effects of the immunosuppressant such as nephrotoxicity, malignancies or infections play a role. Several studies revealed a positive effect of the m-TOR inhibitor everolimus (Eve) on CAV and renal function. However there are still limited data in children and adolescents. This study was performed to assess the effects on CAV and some side effects as renal function, cardiovascular risk factors, post-transplant lymphoproliferative disorder (PTLD), acute rejections and CMV infections after conversion to Eve in heart transplanted children / adolescents and to gain initial insights into this novel treatment regimen. Additionally the conversion reasons were analysed.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Methods: In this retrospective, single-center study 36 patients were switched to everolimus and observed for up to four years, and descriptive before-and-after comparisons were performed.
Results: 36 patients (mean time after transplantation 6.3 ± 4.7 years) were recruited. Indications to conversion were CAV in 33.0% and renal insufficiency in 16.7%. In terms of the CAV among 9 patients, four showed no progression, three an improvement, one a deterioration of a CAV and one patient a first diagnosis. Renal function shows different courses. The mean kreatinin-clearance of 16 patients at 6 months increased from 95.5 ± 25.8 ml / min / 1.73m^2 (range: 43.5-155.4) to 99.6 ± 32.9 ml / min / 1.73m^2 (range: 49.1-185.0). There were trends in increasing lipid levels and decreasing blood glucose levels. In three cases of PTLD the symptoms disappeared and no new event occurred. There were no acute rejections or CMV infection.
Conclusions: In children and adolescents a switch to everolimus is a safe immunosuppressive regimen without increasing the risk of acute rejection or CMV infection. Renal function appears to be improved, with different courses overall. There was a trend to reduce the progression of the CAV however there was a risk of an increase in lipid parameters. In addition there were indicators of a positive influence on glucose metabolism including post-transplant diabetes, as well as a PTLD.

O8-1
Obstetrical and neonatal outcomes in pregnancies with prenatal diagnosis of fetal congenital heart disease: a multicenter experience

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Introduction: A fast-increasing number of studies have paved the way for a possible link between an impared placenta's function and pregnancies affected by congenital heart disease (CHD). Hypertensive pregnancy disorders (HDP) that are most common placenta-related complications put mother and fetus at an increased risk of morbidity and/or mortality. The aim of this retrospective study was to explore the incidence of HDP and preeclampsia (PE) in pregnancies affected by CHD.
Materials and methods: Patients included into the study were referred to a tertiary Fetal Cardiology Center from 2003 to May 2018. Out of 1043 pregnant patients with offsprings affected by CHD, after excluding terminations of pregnancy, intrauterine fetal deaths, twin pregnancies and chromosomal abnormalities, 480 cases were studied for HPD and PE. Outcomes of CHD group were compared with those of a control group without CHD (456 cases followed in the Dpt. of Obstetrics). Univariated analysis by Fisher's/Chi-square tests and adjusted binary outcome values by logistic regression were performed. Moreover, inter-group comparisons among different CHD were made by Kruskal Wallis test.
Results: CHD pregnancies showed placental-related complications like PE (14/480, 2.9% vs 4/456, 0.9%), HDP (21/480, 4.3% vs 15/456, 3.3%) and placental abnormalities (22/480, 4.5% vs 15/456, 3.3%) more frequently than control group, but statistical significance difference was achieved only for PE (p=0.023). The adjusted OR for maternal factor (age, parity, mode of conception and comorbidity) was significantly increased for both conditions (HDP – OR 2.7 [95 CI: 1.0-6.82], PE – OR4.1 [95CI: 1.1-16.3]. PE and HDP were significantly more frequent in Tetralogy of Fallot /ToF/ (4/50) and 5//50, p<0.05, hypoplastic left heart /HLHS/ (2/19 and 3/18, p<0.05) and heterotaxy syndromes with CHD (PE 2/14 p< 0.05) than in controls spontaneously conceived (1/239 and 3/239).
Conclusions: This study show an increased risk of obstetrical complications such as PE and HDP, in pregnancies affected by CHD, with higher risk in TOF, HLHS and hetorotaxy syndromes. Consequently, an accurate screening and monitoring for placental disorders and preterm birth should be carried out in all pregnancies with fetuses with CHD.

O8-2
Peripheral flows in fetuses with ductal-dependent congenital heart defects

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Introduction: Abnormal flows in fetal middle cerebral artery (MCA) and umbilical artery (UA) are generally associated with placental insufficiency, but they may also be due to changed hemodynamic conditions in various congenital heart defects (CHDs). Our aim was to examine the influence of ductal-dependent CHD type on MCA and UA flows, as well as on the fetal head growth.
Methods: We retrospectively reviewed echocardiograms of normal fetuses (n=1897), cases with hypoplastic left heart syndrome (HLHS; n=373) and CHD associated with pulmonary atresia (PA; n=190). Fetuses with major extracardiac defects, chromosomal abnormalities, small for gestational age, from multiple pregnancy, and of mothers with pregestational diabetes and hypertensive disorders were excluded from the study. We measured fetal head circumference (HC), pulsatility indices (PI) in UA and MCA and cerebroplacental ratio (CPR = MCA PI/ UA PI).
Results: In HLHS fetuses lower MCA PI and CPR and higher UA PI values were observed (p<0.05). In the group with PA, UA PI and CPR values were higher, (p<0.05), and MCA PI were comparable to normal. UA PI was increased more in HLHS than in the PA group. Head circumference was slightly lower, yet within normal limits in HLHS group (borderline significant), with no difference between normal and PA groups.
Conclusions: Observed peripheral flows in HLHS and PA groups are consistent with hemodynamic changes in these CHDs. Increased UA PI values are probably secondary to diastolic steal, which is more pronounced in case of HLHS than in PA (cerebral vascular resistance lower than pulmonary vascular resistance). Cerebral flow is impaired in HLHS both in systole and diastole (filling only retrograde through hypoplastic aortic arch), which gives another reason for decreased MCA PI, and is not the case in PA fetuses. Observed difference in head growth in HLHS fetuses, however mild, may be due to impaired blood flow to the head region.

Group	Normal (n=1897)	HLHS (n=373)	PA (n=190)
Gestational age [weeks]	13-40 (median 26)	13-40 (median 26)	14-40 (median 27)
No of measurements			
MCA PI	1346	348	171
UA PI	1451	356	175
CPR	1333	343	166
HC	1885	366	188

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

O8-3

Prenatal diagnosis of transposition of the great vessels with a restrictive foramen ovale

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Objectives: We analyzed our own experience in the prenatal diagnostics of transposition of great arteries with intact ventricular septum and restrictive foramen ovale and perinatal management for this group of patients.
Methods: We analyzed fetal echocardiograms of patients with a diagnosis of transposition of great arteries with intact ventricular septum at our institution from 2004 to 2017. Based on the echocardiographic findings, we identified predictors of restriction and allocated risk group.
Results: From 2004 to 2017 in our Center a diagnosis of transposition of great arteries with intact ventricular septum in 212 fetuses was established. In 74 cases, during the consultation after 36th weeks, a diagnosis of a restrictive foramen ovale was established. It has been confirmed in 61 (82,4%) patients after birth – 3 (4,9%) patients died just after birth before conducting any interventions. An urgent atrioseptostomy was performed in 10 (16,4%) patients in the delivery room, and in 42 (68,8%) – during the 1st day of life in the the cath lab. The arterial switch operation was performed in 182 cases – in the first hours of life in 89 (48,9%) cases and on average in 6th±4 a day of life in 93 (51,1%) cases. The mortality in this group of patients was 3,8% (n = 7). In all fetal echocardiograms we evaluated the size and anatomy of the oval window, including the opening angle, size and configuration of the aneurysm (if present). The relation in size of all cardiac structures and their Z-score, aortic and pulmonary velocity time integral were measured and analyzed. From September 2016, the routine use of an oxygen test (22 fetuses) was introduced. The maximum and mean velocity on pulmonary veins, left and right ventricular outflow tract, aortic arch and ductus arteriosus velocity time integral was measured before and after the oxygen test.
Conclusions: The restriction of the foramen ovale may be underestimated in the prenatal diagnosis of transposition of large arteries, which directly affects morbidity and mortality before surgical treatment. Restrictive foramen ovale requires careful diagnosis and urgent atrioseptostomy right after birth.

O8-4

Predictors of coarctation of the aorta in left-right asymmetry of the 2nd trimester

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Background: Predicting neonatal coarctation of the aorta in left-right asymmetry is still challenging.
Objectives: To identify absolute cardiac dimensions predicting neonatal coarctation in fetuses diagnosed during the 2nd and who had repeated echoscan during the 3rd trimesters, and if the evolution of measurements and L-R ratios could improve the accuracy to predict coarctation.
Materials and Methods: Over a period of 5 years, all fetuses diagnosed with ventricular and great vessels disproportion diagnosed before 28 weeks, followed during the 3rd trimester (34 weeks), and delivered at our institution were included into the study. All cardiac measurements were expressed in Z-score and L-R ratio were calculated. The postnatal outcome was the presence of a coarctation or not. Logistic regression analysis was used to identify the predictors of coarctation of the aorta.
Results: 109 at-risk fetuses were included of whom 72 (66%) developed coarctation. The Z-score values predicting coarctation were for the absolute values: ascending aorta ≤-1.66, aortic isthmus ≤-2.8, and mitral valve ≤-1.34. The L-R ratios associated with coarctation at 2nd and 3rd trimester echoscans were respectively: TV (tricuspid valve)/MV (mitral valve)>1.46 (p= 0.024) and TV/MV>1.60 (p=0.021); PV (pulmonary valve)/AV (Aortic valve)>1.76 (p=0.004) and PV/AV>1.55 (p<0.001); RV (Right ventricle)/LV (left ventricle)>1.31 (p=0.0015) and RV/LV>1.45 (p=0.24). A growth rate of the aortic isthmus ≤0.1mm/week, of the transverse aortic arch ≤0.05mm/week, and of the MV ≤0.3mm/week were associated with coarctation (p=0.028; p=0.0083 and p=0.0003, respectively). Finally, a delta Z-score between the 3rd and the 2nd trimester for the MV ≤0.88, the LV end-diastolic dimensions ≤-1.83, and the aortic isthmus ≤-1.08 predicted neonatal coarctation (p=0.0015; p=0.0211 and p=0.013, respectively).
Conclusion: A multiple prediction parameters model and dynamic of cardiac dimensions during pregnancy may improve the prediction of neonatal coarctation in 2nd trimester L-R asymmetry.

O8-5

Fetal Cardiac Tumors: Long Term Follow-up

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Introduction: Cardiac tumors are rare in children with an incidence of 0.009% in prenatal evaluations and 0.08 -0.2% in pediatric cardiology clinics. Rhabdomyoma is the most frequent, and although benign, serious conditions might be associated such as hemodynamic compromise, arrhythmias or Tuberous sclerosis and its neurologic compendium. The aim of this study is to describe prenatal and postnatal management and long-term follow-up of fetal cardiac tumors.
Methods: Retrospective study of fetal cardiac tumors diagnosed between 1998-2017 at our fetal cardiology unit. All Clinical records were reviewed: fetal echocardiography, prenatal and postnatal management, postnatal echocardiography and neurological status.
Results: A total of 36 fetuses with cardiac tumors were identified at a mean gestational age of 30 weeks. Seven legal pregnancy interruptions were performed (4 neurological involvement, 1 teratoma with hydrops, 2 intracardiac tumors). Rhabdomyoma represent 90%, 5% fibroma, 2.5% teratoma, 2.5% pericardium tumor. Electrocardiogram abnormalities were found in 17% (1 complete auriculoventricular block with remission at 9 months of age, first degree AV block, 3 supraventricular and ventricular premature beats). Surgery was required in 10%: 1 ventricular outflow obstruction, 1 progressive growth, 1 massive pericardial effusion (despite evacuating pericardiocentesis performed in utero). Median follow-up of the 29 newborns was 8,5 years (1-19 years) with an overall survival of 96,5%. One death was reported secondary to cardiogenic shock in a giant obstructive neonatal tumor prior starting ECMO program (1999). Although most patients are currently cardiologically asymptomatic with spontaneous regression of the tumors without medical treatment, 53% associate tuberous sclerosis of which 58% have serious neurologic manifestations such as West syndrome, autistic spectrum disorder and neurodevelopmental delay. A neonate with giant left ventricle rhabdomyoma with severe hemodynamic compromise had an outstanding response to everolimus.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusions: Prenatal diagnosis of cardiac tumors is standard practice. Although most tumors are benign, they might associate serious neurologic comorbidity or a relevant need of cardiac surgery with extracorporeal circulation and a non-negligible mortality. Everolimus can be considered as a treatment alternative for multiple rhabdomyomas or those that cannot be surgically removed.

O8-6

Evaluation of the Value of Next-Generation Sequencing in a Series of 66 Fetuses with Complex Cardiac Malformations

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Introduction: During ultrasound prenatal screening, the discovery of a complex cardiac malformations leads to cytogenetic tests such as FISH caryotype, standard caryotype and/or aCGH. Because of the availability of Next-Generation Sequencing (NGS), we tested whether NGS could yield information on the etiology of some of these cases.
Methods: After ultrasound discovery of a complex cardiac malformation during pregnancy, a fetal sample was performed after obtaining an informed consent to carry out cytogenetic tests and NGS on a panel of more than 400 genes. DNA was prepared with a customized set of probes (Nimblegen, Roche). Eight DNA samples by run were sequenced on a NextSeq500 (Illumina) with a mid-size flow cell. Sequences were analyzed through a pipeline designed locally (VarAP) able to dectect deletion/duplication of exons/gene (DeCovA). Putative causative variants were confirmed by Sanger sequencing and the segregation of selected variants was performed on family members.
Results: 66 fetal DNA were sequenced. A majority of fetuses had heterotaxy and complex cardiac malformations (40 cases) but 26 cases had a severe heart defect (mainly hypoplastic left heart syndrome) but no heterotaxy. A mutation was found in 12 cases (18%) mainly in heterotaxy cases (10 cases) and rarely in non-heterotaxy cases (2 cases). Altogether, 7 genes had a mutation in heterotaxy patients (DNAI1, GDF1 (2x), LMLN2 (2x), MMP21, NEK2, SHH and ZIC3 (2x)) and 2 genes in non-heterotaxy cases (MYH6, TAB2). In most cases, the mutation was recessive autosomal (DNAI1, GDF1, LMLN2, MMP21, NEK2, MYH6) or X-linked (ZIC3) but in 2 cases the mutation was dominant with very variable expressivity (SHH, TAB2). No de novo mutations were discovered.
Conclusions: Discovering causal gene mutations in fetal cases with complex cardiac malformations is feasible and leads to a specific genetic cause in 25% of fetuses with heterotaxy and less than 8% of cases with no-heterotaxy. Uncovering the genetic cause of fetal cardiac malformation does not change the course of pregnancy which is determined by the cardiac and extracardiac malformations but it allows a genetic counseling to the parents with an accurate evaluation of the recurrence risk for future pregnancies.

O9-1

Congenital mitral stenosis: old problems and new therapeutic options

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Introduction: Congenital mitral stenosis (MS) is a rare but fatal anomaly and its course depends upon the entity of mitral damage and the severity of coexisting lesions. Valve repair could be difficult as the disease may involve the annulus, the leaflets and subvalvar apparatus. On the other hand, valve replacement is burdened by the size of the prosthetic valve. A light at the end of the tunnel could be represented by the use of the melody valve on mitral position. However, this approach is out of label and carries potentially fatal problems. Herein we describe our experience.
Patients and methods: In the last 30 years we operated on 279 patients with a congenital mitral disease. Among them, 40 presented with MS (age 6,9±5,6 years; body weight 16,9±14,8 Kg; 20 females). All patients were evaluated through an echocardiographic examination while a MRI or a cardiac catheterization were adopted to evaluate coexisting anomalies. According to Van Praagh classification, 19 patients had type I MS, none had type II, 12 had type III, finally 9 had type IV.
Results: The valve was repaired in 29 patients using different techniques whose purpose was to remove supramitral ring (14 pts), restore a proper leaflet motion (9 pts) and improve subvalvar apparatus opening (24 pts). Replacement was adopted in 11 patients: 7 received a mechanical valve and 4 had a Melody on mitral position. Overall mortality was 12,5%, but it rose to 45% considering patients younger than 1 year of age. The melody valve was used in patients with the smallest annulus and the mortality rate among them was 50%.
Conclusions: Congenital MS is one of the most deadly congenital heart diseases, and the worst anomaly of the mitral valve. The conservative approach is difficult because of the anatomical disarrangement of the valvar and subvalvar apparatus. Nevertheless, replacement carries detrimental intraoperative and postoperative effects. Recently, surgeons looked kindly to the melody as the best candidate valve. In our experience, this valve suits also in small annulus, however, the results are quiet poor. There is still a way to go before achieving lasting and satisfactory results.

O9-2

Long term survival and re-intervention free survival after surgical correction of complex congenital heart defects in childhood

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Introduction: To evaluate long-term survival and relevant surgical and/or trans-catheter re-intervention free survival after surgical correction of complex congenital heart defects (CHD) in childhood.
Methods: Data obtained from the database of a single nation-wide paediatric cardiac centre between 1977 and 2016 were cross-mapped with the National Death Registry and the National Registry of Cardiovascular Interventions for adults. Survival was determined by death or heart transplantation after corrective surgery and the first relevant re-intervention was considered for event-free survival analysis using the Kaplan-Meier method.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Results: Twenty five-year probability of survival after surgical correction of CHD and re-intervention free survival were as follows: Tetralogy of Fallot repair (848 patients) 92% with 65% freedom from pulmonary valve replacement; arterial switch for transposition of the great arteries (605 patients) 94% with 90% freedom from coronary artery re-intervention and 85% from pulmonary arteries re-intervention; atrioventricular septal defect repair (550 patients) 94% with 89% freedom from mitral valve replacement; total cavopulmonary connection for univentricular heart (338 patients) 89% with 86% freedom from valve replacement and 84% from pulmonary arteries re-intervention; pulmonary atresia with ventricular septal defect (111 patients) 76% with 36% freedom from conduit replacement; arterial trunk repair (92 patients) 69% with 56% freedom from truncal valve replacement and with 5% freedom from conduit replacement.
Conclusions: Recent early results of interventional treatment of CHD in children are excellent, however the probability of re-interventions is considerable after correction of complex heart lesions.

O9-3
Smaller brain volumes at two years of age in patients with hypoplastic left heart syndrome - Impact of surgical approach

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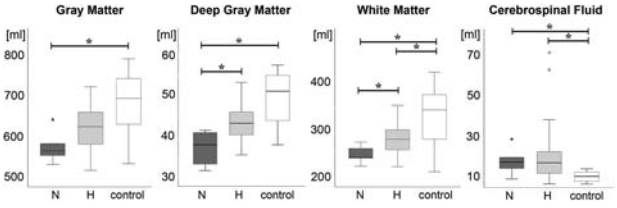
Background: Brain growth in hypoplastic left heart syndrome (HLHS) might be reduced before and after birth. Little is known about further brain growth until two years of age before Fontan procedure and the potential impact of type of surgery.
Methods: In a prospective, two-center study 29 patients with HLHS and variants were treated by Norwood (n=5) or Hybrid procedure (n=24). At two years of age a cerebral MRI was performed and brain volumes (gray, deep gray, white matter) and cerebrospinal fluid were calculated using FreeSurfer image analysis suite and compared to a healthy control group (n=8).
Results: The total brain volumes in patients with HLHS were smaller compared to controls (HLHS: 893 ± 76 mL vs. controls: 1015 ± 148 mL, p=0.005). This difference was found in all three analyzed brain compartments after Norwood procedure, whereas patients after Hybrid procedure had comparable gray and deep gray volumes compared to controls. The reduction of brain matter was more pronounced for deep gray matter (Norwood: 38.4 ± 4.1 ml vs. Hybrid: 44.4 ± 3.9 ml, p=0.005), and white matter (Norwood: 255 ± 19 ml vs. Hybrid: 285 ± 31 ml, p=0.032) for Norwood patients compared to Hybrid.



Conclusions: Smaller total and regional brain volumes were found two years after stage I/II Norwood or Hybrid procedure in children with HLHS. The brain volume reduction was more distinct after Norwood than after Hybrid procedure. Longitudinal studies are needed to identify impact of early staged-surgeries on brain development and may become part of the decision making process in individual patients.

O9-4
Longterm Follow-up After Biventricular Repair of the Hypoplastic Left Heart Complex

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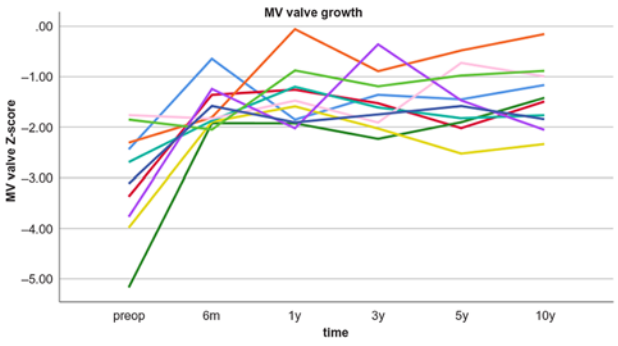
Introduction: In hypoplastic left heart complex (HLHC) patients univentricular palliation can be avoided with more favorable results from biventricular repair. However, studies on patient outcome after biventricular repair are limited and show divergent results. We retrospectively characterized patients with HLHC after biventricular repair, analyzed mid-and longterm outcome and assessed left heart structures growth.
Methods: Patients with HLHC after biventricular repair between 2004 and 2018 were retrospectively reviewed. HLHC was defined as an aortic valve (AoV) or mitral valve (MV) annulus Z-score of less than -2 and a coarctation or aortic arch hypoplasia. Clinical outcome was analyzed and MV and AoV annulus, left ventricular (LV) length and LV internal diastolic diameter (LVIDd) were measured before surgery and 6 months and 1, 3, 5 and 10 years after biventricular repair.
Results: In 30 patients, median age at surgery was 10 days (IQR, 7.75 to 13.25). All patients survived. Fourteen patients (46.7%) required a surgical or catheter based reintervention. Median follow-up was 6.19 years (IQR, 4.12 to 10.16). Mean preoperative Z-scores ± SD (range) were calculated for MV (-2.82 ± 0.96 (-5.17 to -1.10)), AoV (-2.29 ± 1.22 (-4.85 to -0.25)), LV length (-2.09 ± 1.05 (-4.77 to -0.23)) and LVIDd (-2.24 ± 1.61 (-6.45 to 0)). Paired T test showed that most of the catch up growth was seen in the first 6 months after repair, as indicated by the significant difference between mean preoperative and 6 months follow up Z-scores: MV mean difference 1.24, 95% CI: 0.86 to 1.61, AoV mean difference 1.97, 95% CI 1.46 to 2.48, LV length mean difference 1.60, 95% CI 1.12 to 2.09 and LVIDd mean difference 2.70, 95% CI 2.07 to 3.33. At 10 year follow-up all dimensions normalized, with limited growth of the MV (mean Z-score ± SD is -1.41 ± 0.64).



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusions: Biventricular repair in HLHC patients can be performed with good results, although almost half of the patients required a reintervention. Growth of left sided cardiac structures is most important in the first 6 months after repair with Z-scores almost normalized at 10 years follow up.

O9-5

Congenitally corrected transposition of the great arteries: is it really a transposition?

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Introduction: Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital malformation which associates atrioventricular discordance and ventriculo-arterial discordance. Although a ventricular septal defect (VSD) is frequently associated, its anatomy remains controversial. This could be due to the apparently different anatomy of the left-sided right ventricle (RV) compared to a right-sided RV. We wanted to compare the RV septal anatomy between ccTGA, transposition of the great arteries (TGA) and normal heart (NH) and to determine the anatomy of the VSD in ccTGA.

Methods: We analyzed 102 human heart specimens: 31 ccTGA, 36 TGA, 35 NH. According to the last classification of VSD (ICD 11), VSD were classified as outlet if located above the septal insertions of the tricuspid valve, inlet if underneath. We measured the lengths of the anterior (AL) and posterior (PL) limbs of the septal band and the angle between the two limbs. In order to assess the orientation of the septal band, we also measured the angle between AL and the arterial valve above (AL-AV).

Results: VSD was present in 26 ccTGA (83.9%) and was an outlet VSD in 17 cases (65.4%). Mean AL-PL angle was 76.4° for ccTGA compared to 90.6° for TGA (p=0.011) and 76.1° for NH (p=ns). Mean AL-AV was 70.6° for ccTGA compared to 90.6° for TGA (p=0.0004) and 69.1° for NH (p=ns). PL was significantly shorter in ccTGA (p<0.0003): AL/PL length ratio was 21.4 for ccTGA, 2.2 for TGA and 1.5 for NH.

Conclusion: The typical VSD in ccTGA is an outlet VSD. Its frequent misdiagnosis as an inlet VSD is due to the short PL, which creates the illusion of a posterior VSD. Surprisingly, the orientation of the septal band is similar in ccTGA and NH, despite the atrioventricular discordance, and different in ccTGA and TGA, despite the ventriculo-arterial discordance. Even if a transposition is present, ccTGA should not be considered a sub-group of TGA and the term "double discordance" might be more appropriate. These anatomic characteristics may also play a role in the dysfunction of the systemic RV, which occurs earlier in TGA post-atrial switch than in ccTGA.

O9-6

Influence of Echo derived Preoperative Pulmonary Annulus Z value on late outcome of TOF: Outcome of 123 consecutive TOF repairs followed up for 959 patient years

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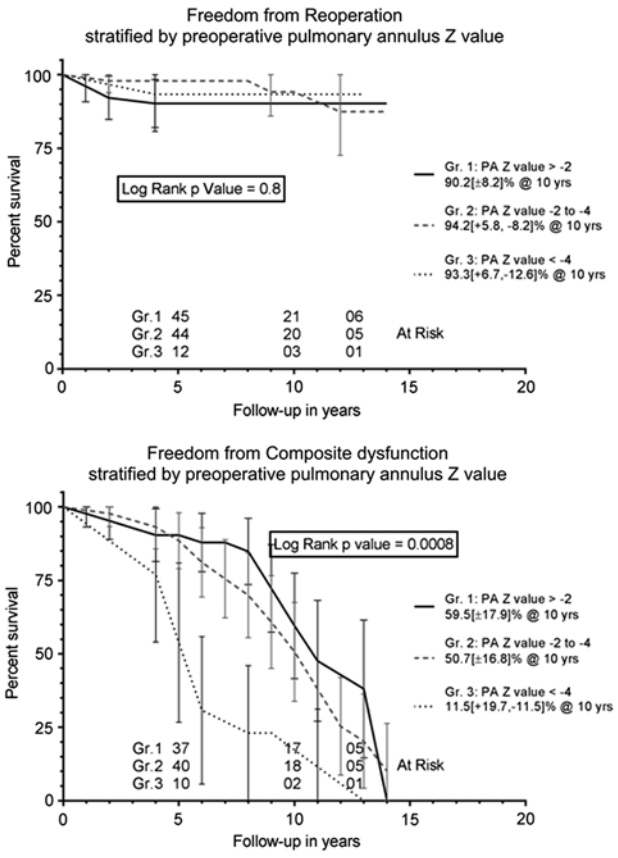
Introduction: Long-term outcome of TOF depends on the size and function of the pulmonary Valve. This study analyzes the impact of preoperative pulmonary Z Score on the long-term outcome of TOF.

Methods: A cohort of 123 consecutive TOF repairs (2004-2014) were divided into Group I [Z > -2] 52; Group II [Z -2 to -4] 54, Group III [Z < -4]17 patients, based on preoperative pulmonary annulus(PA) Z Score. One primary repair using Contegra was excluded from the study. Median age and weight were 161(6-593) days and 5.8(3-8.9)kg respectively. Non-Transannular technique was used in 41(33.3%) patients (Group I:21; Group II:17, Group III:3); Trans-annular technique was used in 82 (66.7%) patients (Group I:31; Group II:37, Group III:14), thus resulting in trans-annular repair in 59.6% of Group I; 68.5% of Group II and 82.4% of Group III patients. Composite pulmonary valve dysfunction was defined as peak gradient > 40 mm Hg and/or pulmonary regurgitation ≥ moderate at follow-up.

Results: All patients were alive at a median of nine(4-14) years of follow-up. 21(17%) patients needed catheter intervention: mostly for branch PA stenosis or for Melody valve insertion. 11(9%) needed reoperations, including for pulmonary valve insertion, tricuspid valve repair and side branch stenosis. A total of 10(8%) patients needed pulmonary bio-prosthesis either surgically (7) or trans-catheterly (3). At last follow-up, freedom from composite pulmonary dysfunction was 59.5±17.9%, 50.5±16.8% and 11.5 [+19.6, -11.5]% @ 10 years for Groups I-III respectively (p=0.0008). Freedom from replacement was not significantly different between groups (p=0.8) [Fig. 1]. Freedom from catheter reintervention was 88.6±9.9%, 81.8±13.1% and 88.2[+11.8, -15.3]% @10 years respectively (p=0.5).



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusions: While the limits of valve sparing approaches are being pushed, we have followed a middle path of relieving obstruction while also achieving pulmonary valve competence. This strategy has yielded excellent freedom from replacement as well as reintervention in the most disadvantaged group with Z > -4. The low freedom from composite dysfunction in Group III suggests the work pending before us to device methods to improve composite function in the long-term.

O10-1
Ventricular dysfunction rather than increased pulmonary vascular resistance is the predominant cause of Fontan failure - Haemodynamic and echocardiographic findings in adults with failing Fontan

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Objectives: Despite its short- and long-term success, the Fontan circulation is palliative and an increasing number of patients will eventually experience Fontan failure during long-term follow-up. Pathophysiology of failing Fontan is complex and conventional treatment options are usually of limited success. Increased pulmonary vascular resistance (PVR) is a frequently incriminated cause of Fontan failure but also ventricular dysfunction is not uncommon. We sought to characterize longitudinal haemodynamic and echocardiographic findings in adult failing Fontan patients to determine the most prevalent causes of haemodynamic failure.
Methods: From the entire cohort of Fontan patients treated in our institution (N=443), adults >18 years of age at last follow-up (n=164) were retrospectively screened for failure (characterized by NYHA III-IV without improvement and/or >2 unscheduled hospitalizations within 12 months and/or active protein-losing enteropathy). Haemodynamic and echocardiographic findings were collected and analysed.
Results: We identified 40 patients (median age 28.5 years [IQR 23.9-36.3], median follow-up after Fontan 21.4 years [IQR 14.8-24.0]). Of these, 29(72.5%) had moderate to severe systolic ventricular dysfunction (ejection fraction ≤45%, n=15) and/or evidence of diastolic dysfunction (end-diastolic ventricular pressure, EDP ≥12mmHg, n=18). Borderline / elevated PVR (calculated index 2-2.5/≥2.5 WU*m^2) was only seen in 5(12.5%) / 7 (17.5%) patients, respectively. Overall, ejection fraction declined significantly from 63±6% at early follow-up (2.1 years [IQR 0.6-10.8]) to 48±14% at last follow-up (p<0.001). Pulmonary artery pressure increased from 11±3 to 15±5 and EDP from 6±3 to 12±5 mmHg (both p<0.001), while transpulmonary gradient (5±2 vs. 4±2 mmHg) and PVR index (2.0±1.1 vs. 1.7±0.8 WU*m^2) did not change significantly (p>0.05). Mortality in adult failing Fontan patients was substantial, 20(50%) died during follow-up.
Conclusions: Therapeutic options in a truly failed Fontan circulation are limited and sustained recompensation by medical, interventional or surgical means is rare. Therapies targeting reduction of PVR may be of limited success since systolic and, rather underappreciated, diastolic ventricular dysfunction seem to be the predominant causes of Fontan failure. Heart transplantation as ultima ratio should be considered timely, before progressive multi-organ dysfunction impedes successful transplantation, especially in the light of donor organ shortage and extended waiting time, since mortality in failing Fontan patients is high.

O10-2
Intima Media Thickness and Cardiovascular Risk Assessment in 764 Adults with Congenital Heart Disease

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Introduction: As long-term survival of adults with congenital heart disease (ACHD) has improved, risk identification for atherosclerotic cardiovascular disease becomes more important in their primary care. Increased intima media thickness (IMT) serves as a marker of structural atherosclerosis and is a valid indicator for future cardiovascular events. This study compared IMT of ACHD with healthy controls and the association with the 10-year risk for a major cardiovascular event (PROCAM score).
Methods: From February 2015 to December 2018, 764 ACHD (356 female, 38.5 ± 11.8 years) and 195 healthy volunteers (114 female, 36.9 ± 13.4 years) received a sonographic assessment of the IMT at the common carotid arteries. In 645 ACHD blood pressure along with information on anthropometrics, smoking habits, cardiovascular family history, and antihypertensive therapy was also taken to calculate the risk of myocardial infarction or stroke within the next 10 years according to the PROCAM risk score.
Results: After adjusting for age, sex, BMI, and current intake of hypertensive drug medication mean IMT of ACHD was 0.538 ± 0.083 mm and ACHD did not show differences to healthy controls (0.541 ± 0.084 mm, p=.684). Only patients with coarctation of the aorta (CoA, n=69) showed a significantly higher IMT (0.590 ± 0.075 mm, p<.001) compared to healthy controls. Furthermore, IMT showed just poor association to the PROCAM risk score (r=.110; p=.005).
Conclusion: IMT was rather normal in ACHD and only increased in patients with CoA. These structural changes of the vasculature were only poorly correlated to the PROCAM 10-year risk evaluated for the general population. Whether both parameters are relevant for cardiovascular risk assessment in ACHD needs to be clarified in further outcome studies.

O10-3
Surgical versus percutaneous stenting treatment of isolated aortic coarctation: long-term follow-up

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Objectives: We sought to compare long-term outcomes of surgical versus percutaneous stenting strategies for aortic coarctation (CoA) repair in a large cohort of pediatric and adult patients.
Methods: We identify 212 patients (20±8,7 yrs, 72% male, 47% <18 yrs of age) with isolated CoA with median follow-up of 17 years after aortic repair. Patients were divided into 3 groups: 139 (median age at repair 39 days, 68% male) with single-time surgical repair (end-to-end anastomosis, patch angioplasty or subclavian flap) (CoA-S group), 18 (median age at repair 12 yrs, 56% male) with single-time percutaneous stenting (CoA-PS group) and 55 (median age at repair 23 days, 85% male) with recurrence of CoA who underwent multiple aortic procedures with or without balloon angioplasty (BA) intervention (CoA-H group). All subjects underwent 24-hour ambulatory blood pressure monitoring (ABPM) and trans-thoracic echocardiography.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Results: HTN therapy was observed in a significantly higher proportion (83%) of patients in the CoA-PS group, compared to 65% and 46% of CoA-H and CoA-S patients, respectively (p=0.002). Also, CoA-PS group exhibited a significantly higher proportion of mean daytime systolic BP values above 95th centile at ABPM (22% vs 6,5%, p=0.045). Echocardiogram revealed a higher median residual aortic gradient at the site of repair in CoA-H compared to CoA-S patients (p<0.001) and the number of patients with re-CoA gradient >20 mmHg was progressively higher in CoA-S (33%), CoA-PS (50%) and CoA-H (73%) groups (p=<0.0001). At Kaplan Meier survival analysis, stent treatment was associated with significantly higher incidence of recoartaction (log rank p<0.0001) compared to other techniques. At multivariate regression Cox analysis adjusted for gender, age at CoA repair, BMI>90° and need for HTN therapy, aortic stenting was an indipendent predictor of echocardiographic evidence of recoarctation (H.R. 7,931, 95% CI 2,870-21,916 p=<0.0001).
Conclusions: In our study, CoA stenting was independently associated with recoartaction during long-term follow up when compared to surgical procedures. Furthermore, patients with percutaneous stenting had lower blood pressure control at ABPM and higher need for antihypertensive therapy.

O10-4
Molecular signaling pathways in right heart failure of adult patients after tetralogy of Fallot repair
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Introduction: Right heart failure (RHF) secondary to pressure and/or volume-overload contributes to significant morbidity and mortality in the growing population of adult patients after tetralogy of Fallot (TOF) repair. The goal of this study was to describe signaling pathways contributing to right ventricular (RV) remodeling by analyzing longitudinal over lifetime alteration of RV gene expression in affected patients.
Methods: RV tissue was collected at the time of cardiac surgery in 13 patients with a diagnosis of TOF. RNA was isolated and whole transcriptome sequencing was performed. Gene profiles were compared between a group of 6 adults with signs of RHF undergoing RV-PA conduit surgery and a group of 7 infants undergoing elective TOF correction. Definition of RHF was based on clinical symptoms, such as fluid retention, dyspnea and/or arrhythmia, and evidence of RV hypertrophy, dilation, dysfunction or elevated pressure on echocardiographic, cardiovascular magnetic resonance, or catheterization evaluation.
Results: Median age was 34 years (range 30 – 62 years) in RHF patients and 5 months (range 4.8 – 5.8 months) in infants. Based on an adjusted p-value of less than 0.001, RNA sequencing of RV specimens identified a total of 1927 differentially expressed genes in adult patients with TOF and RHF as compared to infant patients with TOF and no RHF. Gene Ontology and Kyoto Encyclopedia of Genes (KEGG) databases highlighted pathways involved in cellular metabolism, cell-cell communication, cell cycling and cellular contractility to be dysregulated in adult patients with corrected TOF and chronic RHF. Relevant genes and pathways are depicted in the figure.
Conclusions: Right ventricular transcriptome profiling in adult patients with RHF after TOF repair allows identification of signaling pathways contributing to pathologic RV remodeling and helps in the discovery of biomarkers for disease progression and of new therapeutic targets.

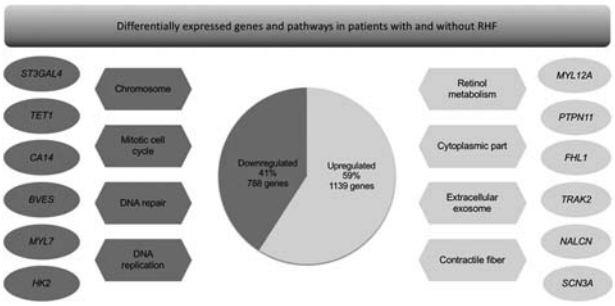

Figure.

O10-5
Cognitive and cerebral MRI findings in a GUCH population treated in the late '90s
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Introduction: Infants treated for congenital heart disease (CHD) by invasive procedure including cardiovascular surgery have an increased risk for impaired neurocognitive function, which is well known for the pediatric age group. In contrast, little is known in the growing up congenital heart (GUCH) population. The aim of this study was to evaluate neurocognitive abilities in a group of young adults treated in the late '90s and their possible association with structural magnetic resonance imaging (MRI) findings.
Methods: Prospective cohort study on GUCH population recruited from the University Heart Center in Zurich and compared to healthy peers. Intelligence quotient (IQ) was determined as part of an extended neurocognitive test battery using the vocabulary and matrix reasoning subtests from the Wechsler Adult Intelligence Scale, Forth Edition (WAIS-IV). Information about socioeconomic background, disability and health status of the patients was collected by questionnaire. Cerebral MRI was performed on a 3T GE MR750 scanner and inspection of any abnormalities was done blinded.
Results: Mean (range) age of 68 enrolled GUCH patients (46% females) and 55 peer controls (49% females) was 26.9 years (19.2-32.7) in the GUCH and 26.4 years (19.9-32.6) in controls. Mean IQ was 98.8 (68-123) in GUCH population and 104.5 (77-129) in controls (95%-CI: -10.67 to -2.12, p = 0.0037). Complexity of CHD had an influence on IQ ($F(2,63) = 3.87$, $p = 0.026$) whereas those with a severe CHD performed significantly worse compared to those with a moderate CHD (mean IQ 90.7 versus 101.0, 95%-CI: -19.036261 to -1.2934093, p = 0.021). Cerebral MRI could be obtained in 47 of the 68 GUCH patients (69.1%) and in 54 of 55 controls (98.2%). Abnormalities on cerebral MRI were discov-ered in 63.6% of GUCH and in 3.0% of

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

controls (p<0.0001). They consisted of focal infarction or at-rophy, microhemorrhages, enlarged cerebrospinal fluid space and abnormal T2 hyperintensities. There was no difference in IQ between patients with or without abnormalities on brain MRI (mean IQ 96.6 versus 100.4, 95%-CI: -2.73 to 10.29, p = 0.25).
Conclusions: GUCH patients are at increased risk of cognitive impairment with a high prevalence of structural cerebral MRI findings.

O10-6
Inspiratory Muscle Training improves Oxygen Saturation and Hemoglobin Levels in Patients with Fontan Circulation – Results from a Randomized Home-Based Training Study

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Introduction: Pulmonary blood flow in patients with Fontan surgery is mainly driven by left heart suction forces and negative intra-thoracic pressures during inspiration. Many patients suffer from decreased oxygen saturation (SpO_2) due to inhomogeneous lung perfusion or venovenous fistulae. Lower SpO_2 leads to elevated hemoglobin (Hb) levels for compensation. This study investigated, whether a daily, home-based inspiratory muscle training (InMT) can influence SpO_2, Hb and peak oxygen uptake (VO_2peak) in adult patients with Fontan circulation.
Methods: 39 patients (female: 46%; 30.6 ± 8.2 years; BMI: 23.5 ± 4.4) with Fontan circulation were randomized into either an intervention (IG) or control group (CG). The IG (n=18) performed a telephone-supervised, daily InMT of 3 sets with 10-30 repetitions for six months with an inspiratory resistive training device (POWERbreathe). Patients randomized into CG (n=21) continued their usual activities. At baseline and final evaluation Hb was determined from peripheral venous blood and SpO_2 was captured by pulse oximetry at rest and during a cardiopulmonary exercise test (CPET). Data from the IG was compared to the data from the CG with a Wilcoxon rank sum test. All values are displayed in median and interquartile [IQR 25; 75].
Results: After six months of InMT, SpO_2 at rest increased in the IG in comparison to a slight decrease in the CG (delta SpO_2 at rest: IG: 1.50 [-0.25; 3.00] % vs. CG: -0.50 [-1.75; 0.75] %; p=.017). Hb level decreased in the IG compared to an increase in the CG (delta Hb: IG: -0.20 [-0.90; 0.20] g/dl vs. CG: 0.40 [-0.25; 0.80] g/dl; p=.040). There was no difference in VO_2peak and SpO2 at peak exercise between both groups (delta VO_2peak: IG: 0.05 [-1.53; 1.33] ml/kg/min vs. CG: -0.50 [-1.20; 0.78] ml/kg/min; p=.784; delta SpO_2 at peak exercise: IG: 1.00 [-2.00; 3.00] % vs. CG: -0.50 [-2.00; 2.00] %; p=.517).
Conclusions: Six months of a telephone-supervised, daily InMT do not affect exercise capacity in patients with Fontan circulation, but improve oxygen saturation at rest and consecutively reduces the primarily elevated hemoglobin level.

O11-1
Impaired biventricular filling and response to percutaneous pulmonary valve implantation in patients with RVOT dysfunction

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Objectives: Atrial dysfunction and impaired ventricular filling have been described in patients with right ventricular outflow tract (RVOT) dysfunction. While the effect of percutaneous pulmonary valve implantation (PPVI) on ventricular properties has been described, its effect on atrial dimensions and function is less well characterized. We thought to assess left and right atrial volume and function in patients with RVOT dysfunction and to study the impact of PPVI on atrial remodelling.
Methods: Patients with RVOT conduit dysfunction who underwent CMR before and after PPVI as part of their routine clinical assessment were included. Right (RA) and left atrial (LA) end-diastolic (EDV) and end-systolic (ESV) volumes as well as atrial passive and active emptying function were assessed using standard axial cine slices.
Results: One-hundred and eleven patients were included (median age at PPVI 18.6; 6.2 - 53.2 years, 41 females (36.9%), CMR 6.2 ±2.1 months post PPVI). Baseline RA passive emptying function as a correlate for early diastolic ventricular filling showed a significant relationship with invasive RV end-diastolic pressures (r=0.27, p=0.01). Both RA passive emptying function (r=0.23, p=0.04) and LA passive emptying function (r=0.26, p=0.02) were significantly related to peak VO2. After PPVI there was a significant decrease in RAEDV (38±17 to 33±15 ml/m2; p< 0.0001) and RAESV (58±21 to 52±20 ml/m2; p< 0.0001). RA passive emptying function improved significantly (19±6 to 22±6%; p< 0.0001) whereas RA active emptying function decreased significantly (33±14 to 32±10%; p=0.005). LAESV (36±10 to 38±10 ml/m2; p=0.001) but not LAEDV (19±7 to 19±6 ml/m2; p=0.09) increased while LA passive emptying function improved (31±7 to 33±8%; p< 0.001) significantly and LA active emptying function (43±13 to 42±11%; p=0.79) remained unchanged.
Conclusions: RA and LA passive emptying function as a correlate of ventricular diastolic function were significantly related to exercise capacity in patients with RVOT dysfunction. Both improved after PPVI suggesting a positive impact of the percutaneous intervention on diastolic dysfunction in these patients.

O11-2
3D geometry of coronary arteries to predict late clinical events after the arterial switch for transposition of the great arteries

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Background: Predicting late coronary events after the arterial switch operation (ASO) for TGA remains challenging. Screening all patients for late coronary obstruction is questionable in patients with no pre- or per-operative risk factors. Acquired coronary anatomy after the transfer during ASO might be a predictor of coronary events.
Objective: To describe 3D geometric characteristics of the acquired coronary anatomy after ASO for TGA and to identify acquired courses of coronary artery associated with coronary events.
Method: We retrospectively reviewed coronary CT performed at a median age of 5 years after ASO: 100 were from random free-from-coronary-event patients and 21 were from asymptomatic patients who had a coronary event during follow-up (5 right coronary artery, 16 left coronary artery). Using 3D modeling software, we defined and measured 6 geometric criteria for each coronary artery: the clockwise position of the ostium in the aortic root, the angle of coronary stem's first centimeter with the ostium, the height of reimplantation of the coronary ostium, the distance separating coronary ostium and first centimeter from pulmonary artery, and the coronary minimal angulation with the aortic wall.

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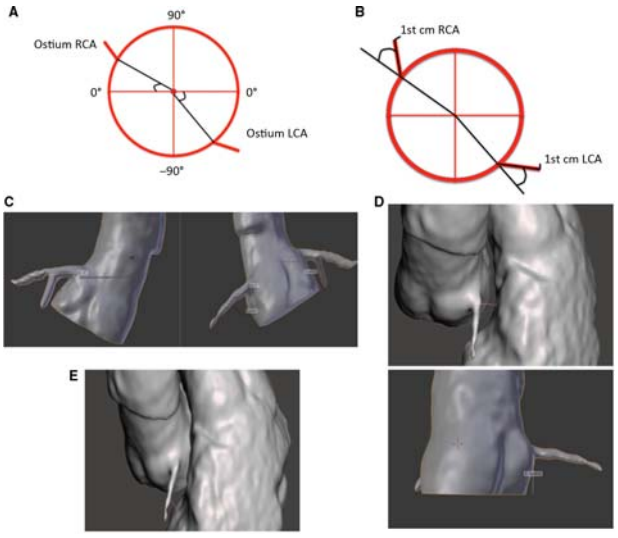

Figure.

Results: None of the geometric parameters studied for the right ostium was associated with coronary events. Four/6 criteria measured on left coronary artery were statistically associated to coronary events: the clockwise position of the left ostium > 67° (p<0.001) (A), the minimal angle with the aortic wall in 3D <39°, p=0.003 (B), the angle of coronary stem first centimeter with the ostium >62°, p<0.01 (C), the distance separating coronary ostium from pulmonary artery <10 mm, p=0.03 (D).
When combining the geometric characteristics, the association of anterior position of the left ostium > 67° and minimal angle in 3D < 39° had a 88% sensitivity and a 81% specificity to predict coronary events (ROC curve; 0.847, IC 95% (0,74; 0,95)), p<0,001.
Conclusion: Efficiency of screening for coronary anomalies after the ASO is limited due to the rarity of late events. Imaging 3D acquired anatomy of coronary arteries might however be useful to select patients at risk of events and to tailor follow-up.

O11-3
Longitudinal analysis of exercise performance in patients after TOF repair

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Introduction: With the increase in survival of Tetralogy of Fallot patients (TOF), treatment strategies may shift their focus from preventing mortality towards improvement of quality of life. A diagnostic tool to early recognize deterioration is the cardiopulmonary exercise test (CPET). Serial CPETs in these patients have been an emerging strategy for therapeutic interventions and monitoring their health status. The purpose of this study is to show the change in exercise performance over time in TOF patients.
Methods: This retrospective cohort study included all TOF patients followed in the German Heart Center of Munich, who underwent a CPET between September 2001 and June 2015. CPETs were included when the respiratory exchange ratio was above 1.00. There were no restrictions concerning the interval between the CPETs. Predictive values were calculated based on Bongers 2014 for children, extrapolated to the age of 25, and Gläser 2010 for adults. The main outcome was peak oxygen uptake as percentage of predicted (peak VO2). Statistical analyses were based on time-dependent growth models.

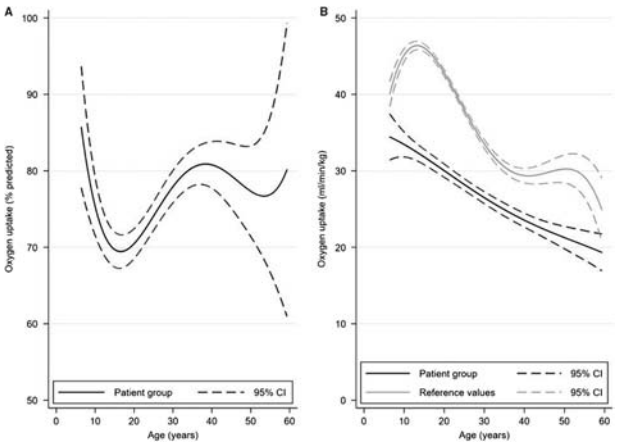

Figure 1.
Exercise capacity over time in 451 individual patients with 1133 measurements expressed as percentage of predicted (A) and as oxygen uptake indexed for body weight (B).

Results: A total of 1133 CPETs were included, corresponding to 451 individual patients. Serial testing of at least 3 follow-up moments was completed in 178 patients. The average age at the first visit was 25.7 ± 11.5, ranging from 6.4 to 63.8 years of age. The interval between CPETs varied from 1 day to 135 months. The peak VO2 at the first visit was 27.7 ± 8.6 ml/min/kg, corresponding to 74 ± 19%. The time-dependent growth model showed a decrease in peak VO2 predicted early in the 2nd decade and an upsloping pattern towards 40 years-of-age (Graph A). This might be in part an effect of the increase of the reference values during puberty, that was not observed in patients with TOF (Graph B). The second decline in exercise performance might be a supra-physiological decline while aging.
Conclusions: These results of serial CPETs in a large cohort of patients with repaired TOF show a pattern of continuous deterioration, whereas the reference population is increasing their exercise capacity during adolescence. Further analyses are warranted to identify modifiers of exercise performance such as interventions, medication use, and comorbidities.

O11-4
Multimodal assessment of vascular function in patients with Fontan circulation

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Introduction: Arterial stiffening is a well-established marker of cardiovascular morbidity in the general population, but there is a relative paucity of data on patients with a functional single ventricle following Fontan palliation. However, due to the unique challenges and long term complications associated with Fontan physiology, understanding vascular function in this population appears particularly important. The aim of this study is to

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

characterize vascular function in Fontan patients using a multimodal approach.
Methods: Patients with Fontan physiology as well as healthy controls were included in this prospective observational study. Central blood pressure (BP), central augmentation index corrected to a heart rate of 75/minute (AIx75) and carotid-femoral pulse wave velocity (PWV) were determined using SphygmoCor XCEL (AtCor). Digital Acceleration Plethysmography (Meridian) was used to obtain an aging index (AI). The reactive hyperemia index (RHI), marker of endothelial function, was determined using EndoPAT (Itamar). Carotid intima-media thickness (cIMT) and distensibility were measured with ultrasound. Laser Doppler (Perimed) with iontophoresis of Acetylcholine (ACh) and Sodium Nitroprusside (SNP) was used to assess endothelium dependent versus independent changes in microcirculation. For statistical analyses Mann-Whitney U and Chi-square tests were used as appropriate to assess difference between groups.
Results: 24 Fontan patients and 24 controls were included in the study. Median age 16, range 7-33 years. There was no significant difference between groups for age, height, weight, gender, heart rate and brachial BP. Central systolic but not diastolic BP was higher in patients versus controls (p=0.029). AIx75 (12,5 [IQR 4,3-17,6] vs -6,8 [IQR -14,5 - 0,88]) and AI (-0,596 [-0,67 – (-0,48)] vs -0,82 [-1,02 - (-0,62)]) were higher in patients versus controls (p<0.001). There was no significant difference in PWV, cIMT, carotid distensibility, microvascular perfusion or RHI between groups.
Conclusion: Fontan patients have abnormal wave reflection (AIx75 and AI) which is already apparent in childhood. This study demonstrates no significant abnormalities in cIMT, carotid distensibility, generalized large artery stiffness, microvascular or endothelial function. Future clinical trials should focus on pharmacologic optimization of AIx75, AI and central blood pressure in Fontan patients.

O11-5
Biventricular function in Ebstein Patients. Intermodality feasibility and reproducibility between feature tracking cardiac magnetic resonance and speckle tracking echocardiography

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Objectives: Quantification of biventricular function in Ebstein anomaly (EA) is challenging, due to the different degrees of displacement of the tricuspid valve (TV), correct segmentation of the functional right ventricle (fRV) and often abnormal geometry of left ventricle (LV) squeezed by the enlarged RV. Speckle tracking (ST) echo has been validated in literature, also for right ventricle. Feature tracking (FT) cardiac magnetic resonance (CMR) has recently been introduced for functional evaluation in complex congenital diseases affecting the right ventricle. We sought to compare the strain values obtained by FT CMR versus ST echo as the gold standard.
Methods: Ebstein patients with and without TV reconstruction prospectively underwent ST echo and FT CMR in one core lab, when possible at the same day. Intermodality differences were assessed using correlation coefficient (Pearson r) and Bland-Altmann analysis (% mean difference ± standard deviation; 95% limits of agreement).
Results: Seventeen patients underwent ST echo and FT CMR. FT analysis of biventricular function was feasible in all patients both for longitudinal (LGS) and circumferential global strain (CGS). Because of bad quality image, ST was feasible in 100%, 64% and 76% of the patients, respectively for left ventricle (LV) GLS, LV GCS and fRV GLS. Despite some agreement was found for fRV GLS between FT and SP (r = 0.6) the mean difference was significant and limits of agreement wide (14.8 ±29.57; -43/72). For LV GLS and GCS measurements by FT and ST differed even more (7 ±31; -68/54 and 22 ± 45; -67/111) with poor correlation for both (r=0.3).
Conclusions: FT CMR is more feasible than ST echo in Ebstein patients, due to better image quality and the complex geometry of the heart in EA. FT measurements and echi value show wide limit of agreements, meaning that these two techniques are not interchangeable. Therefore follow up assessment of Ebstein patients should be consistently performed by using the same modality.

O11-6
Diffuse myocardial fibrosis is associated with type of adverse loading and location of the right ventricle in congenital heart disease – a cardiovascular magnetic resonance T1 mapping study

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Introduction: While focal myocardial fibrosis is commonly detected by late gadolinium enhancement cardiovascular magnetic resonance (CMR) and has been related to adverse outcome in various congenital heart disease (CHD), the extent of diffuse myocardial fibrosis and its role in CHD are relatively unknown. This study sought to assess myocardial extracellular volume (ECV) reflecting diffuse myocardial fibrosis, and to investigate associations with clinical and functional parameters, type of adverse loading and location of the right ventricle (RV) in CHD.
Methods: CHD patients (n=53, median age 26.1 years) with pressure and/or volume overload of the RV were prospectively enrolled and compared to healthy controls (n=19, 24.8 years). Participants received standardized CMR, laboratory and cardiopulmonary exercise testing. Modified Look-Locker Inversion recovery (MOLLI) T1 mapping was performed in midventricular short axis and axial orientation to quantify RV and left ventricular (LV) ECV.
Results: RV and LV ECV were significantly higher in patients (median 31 and 28%) than controls (29 and 25%; p=0.021, respectively), with values above the upper limit of normal in 32 and 25% of the patients. Within the patient group, LV ECV correlated with N-terminal pro brain natriuretic peptide (r=0.61, p<0.001) and indexed LV stroke volume (r=-0.37, p=0.017). Increased RV ECV was related to volume overload (p=0.024), while greater LV ECV was associated with pressure overload of the RV in systemic vs. subpulmonary location (p<0.001).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusions: CHD patients had higher RV ECV as well as LV ECV in correlation with markers of heart failure, indicating an adverse ventricular interaction. Elevated RV and LV ECV were associated with the type of adverse loading and the location of the RV. Further studies are necessary to explore the clinical implications of diffuse myocardial fibrosis, and to assess the utility of non-invasive ECV measurements in supporting risk stratification and guiding treatment in CHD.

O12-1

Cardiac phenotype and genetic variabilities? An example from beta-thalassemia patients

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Introduction: New Echocardiographic modalities such as speckle tracking echocardiography (STE) have not only allowed the early detection of myocardial dysfunction in systemic diseases but have allowed as well to distinguish the pattern cardiac affection. Several mutations are characterized within the beta-thalassemia patients.
Methods: 40 beta thalassemia patients were studied pertaining to two groups, Group 1 with thalassemia causing mutation IVSI (Intervening Sequence)-110 and Group 2 with predominant IVSI-6 mutation. Both groups were subjected to cardiac MRI for measurement of cardiac iron load, 2D speckle tracking echocardiography for measurement of global longitudinal strain (GLS) and its endocardial vs. epicardial component
Results: Group 1 had more cardiac iron load compared to Group 2 (G1:14.1±1.4 vs. G2: 26±2.2, P<0.01), Group 2 had more endocardial longitudinal strain involvement than Group 1 (G1: 20.1 ±3.2 vs. G2: 12±1.1, P<0.01). Epicardial longitudinal strain was more affected in Group 1 compared to Group 2 (G1:15.2±0.9 vs. G2: 17±3.2, P=0.04).Epicardial GLS was well correlated with myocardial iron load (r=78%; P<0.001). Epicardial GLS proved 80% sensitivity in predicting IVSI-110 mutation, seemingly Endocardial GLS has recorded 82% sensitivity in predicting IVSI-6 mutation.
Conclusions: The aforementioned findings point towards different mechanisms of myocardial injury through different genotypes of Thalassemia, Group 1 develops mainly superficial subepicardial dysfunction due to iron load in the more vascularized layers while Group 2 seem to have more involvement of ischemic Subendocardium by unknown mechanisms. This study underlies as well the possibility of using STE in indirect genetic diagnosis of as displayed in this study.

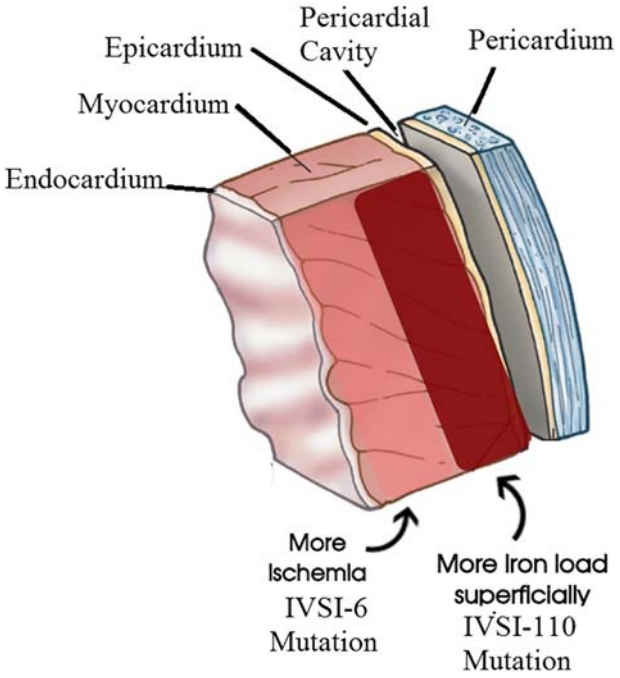

Figure.

O12-2

Study of the time-relationship of the mechano-electrical interaction in an animal model of tetralogy of Fallot: Implications for the risk assessment of ventricular arrhythmia

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Background: Long-term outcome of tetralogy of Fallot(TOF) is determined by progressive RV dysfunction, due to chronic pulmonary regurgitation(PR), and the risk of sudden death by ventricular arrhythmia. Although the electro-mechanical interaction is well-known, its time-relationship remains ill-defined.
Methods: According to contemporary surgical repair, PR was induced by transannular patch with limited RV scarring, in pigs of 25 kg. Biventricular mechano-electrical assessment was based on presuure-volume loops, after respectively 3 (n=8) and 6 (n=7) months, and compared to control animals (n=5). Electrophysiological testing included registration of endocardial monophasic action potentials(MAP) at 3 RV locations (inlet-apex-outflow) and 2 LV locations (septal-lateral), intraventricular conduction velocity, and induction of ventricular arrhythmia based on a burst pacing protocol (minimal cycle length 200 ms).
Results: Data are shown in the table. RV dilation and dysfunction is observed at 3 months, and progressed significantly at 6 months, achieving at that time the critical threshold for pulmonary valve implantation. Depressed RV contractility is associated with impaired LV contractility at 6 months. According to an increased QRS duration, the MAP duration and MAP dispersion at RV and LV were prolonged at 6 months. The RV conduction velocity decreased significantly at 6 months, compared to control and 3 month animals. This was associated with an increased activation time on the 3 RV locations at 6 months. The delayed intraventricular LV conduction was not significant. No sustained ventricular arrhythmias were induced.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Hemodynamics	Control (n=5)	3 months (n=8)	6 months (n=7)	p-value ANOVA
RVEDVi (ml/m^2)	59 ± 16	122 ± 18	142 ± 13	<0,001
RVESVi (ml/m^2)	27 ± 10	74 ± 15	96 ± 7	<0,001
RVEF (%)	55 ± 6*	39 ± 6	32 ± 5	<0,001
LVEDVi (ml/m^2)	78 ± 10	85 ± 17	58 ± 6	0,002
LVESVi (ml/m^2)	36 ± 8	43 ± 10	36 ± 10	0,331
LVEF (%)	54 ± 9	49 ± 10	39 ± 15	0,102
QRS duration (ms)	76 ± 12	88 ± 12	97 ± 13	0,039
MAP duration (ms)				
RV-inlet	260 ± 5	268 ± 12	313 ± 24	<0,001
RV-apex	253 ± 13	262 ± 26	296 ± 37	0,037
RV-outflow	252 ± 21	264 ± 29	308 ± 30	0,005
LV-septal	249 ± 24	274 ± 22	321 ± 43	0,003
LV-lateral	248 ± 12	265 ± 22	327 ± 49	0,001
Activatie time (ms)				
RV-inlet	16 ± 12	33 ± 10	42 ± 10	0,011
RV-apex	7 ± 4	18 ± 6	21 ± 5	0,005
RV-outflow	16 ± 7	36 ± 9	40 ± 13	0,005
LV-septal	5 ± 4	19 ± 10	26 ± 16	0,063
LV-lateral	6 ± 8	25 ± 12	27 ± 16	0,077
Conduction velocity(m/s)				
RV	2,4 ± 0,6	2,1 ± 0,2	1,8 ± 0,2	0,035
LV	2,4 ± 0,7	2,3 ± 0,4	1,9 ± 0,2	0,107

Conclusion: Progressive RV dysfunction is associated with altered electrical properties, observed at 6 months of pulmonary regurgitation, and predominantly affecting the RV. Despite significant RV dilation and concomitant electrical disturbance, ventricular arrhythmia were not induced. The data suggest that the hemodynamical RV deterioration after TOF repair is effectively preceding the risk of inducing sustained ventricular arrhythmia, questioning the need for electrophysiological testing preoperative to pulmonary valve implantation, when extensive RV scarring is absent.

O12-3
Interferon activity in newborns exposed to Ro/SSA autoantibodies in utero and the risk of autoimmune congenital heart block

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Introduction: Autoimmune congenital heart block (CHB) is a rare cardiovascular manifestation of neonatal lupus syndrome. CHB develops in utero in fetuses of women with anti-Ro/SSA autoantibodies. Women carrying Ro/SSA autoantibodies are commonly diagnosed with Sjögren's syndrome or SLE. The maternal Ro/SSA autoantibodies are transferred over the placenta and initiate inflammatory processes, resulting in fibrosis and calcification of the atrioventricular (AV) node and a third degree AVB. Sjögren's syndrome and autoantibody positivity is often concurrent with increased systemic interferon (IFN) activity and expression of IFN regulated genes, with unknown impact on CHB development. Treatment that alters type I IFN activity has been suggested to reduce the risk of CHB. The pattern and magnitude of IFN pathway activation in Ro/SSA autoantibody exposed newborns has not been investigated.
Methods: Thirteen Ro/SSA autoantibody positive mothers either receiving no medication (Ro/SSA+) or treated with hydroxychloroquine (HCQ) and/or azathioprine (Ro/SSA+T), and their newborn babies were included in the study, together with 8 healthy mother-baby pairs (HC). Blood was drawn from the mother and baby (cord) at birth, with immediate separation into plasma and cells. Cellular mRNA expression levels were measured using microarrays and used for calculating IFN scores. Cell surface expression of molecules was investigated by flow cytometry, and IFN-α in plasma and supernatants was analyzed by immunoassays. The ability of immune cells from newborns to produce IFN-α in response to autoantibody exposure was tested by in vitro stimulation assays.
Results: Mothers with Ro/SSA autoantibodies had an increased IFN activity compared to controls, with a higher IFN score and plasma IFN-α levels, which were similarly high in mothers receiving immunomodulatory treatment. Autoantibody-exposed newborns also had significantly increased IFN activation in terms of IFN score and plasma IFN-α levels. The IFN score in newborns correlated positively with the maternal IFN scores in untreated mothers. However, maternal treatment reduced the IFN activation in the babies. Immune cells from newborns responded to autoantibodies by producing IFN-α in culture.
Conclusions: There is an activation of the type I IFN system in Ro/SSA autoantibody exposed newborns which may contribute to the risk of CHB. Immunomodulatory treatment might reduce that risk.

O12-4
Neuregulin-1 (NRG1)-effects on systolic and diastolic function in experimental early childhood RV pressure overload

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Introduction: Pressure load (PL) is the main cause of right ventricular failure (RVF). Functionally, RVF is characterized by progressive diastolic dysfunction. Neuregulin-1, a stimulator of cellular proliferation signaling, has been shown to improve survival in experimental models of PL-induced RVF. Whereas biomolecular data on the cardiac effects of NRG1 are accumulating, the functional effects on ventricular hemodynamics (under persisting pressure load) are unknown. We characterized the effects of NRG1 on systolic and diastolic RV function in an experimental model of early childhood (fixed) RV PL.
Methods: Rat pups (aged 21 days, weighing 30-40 grams) were subjected to pulmonary artery banding (PAB, n=35) or sham surgery (n=18). NRG1 (n=19) or vehicle (VEH, n=16) was injected intraperitoneally from day 3 until day 14 post-surgery. Rats were evaluated daily for clinical symptoms of RVF (e.g. inactivity, cachexia, dyspnea, pleural effusion/ascites). 14 and 28 days post-surgery echocardiography and RV pressure-volume analysis were performed in subsets of the groups.
Results: PAB induced severe PL (all data in table 1). Survival was 100% and merely at day 28 1 rat (in the VEH group) displayed signs of clinical RV failure. In this pre-failure situation we saw that

Table 1 all data ± SEM

	14 days		28 days	
	VEH	NRG1	VEH	NRG1
RV systolic pressure (mmHg)	58±5	62±5	94±7	78±10
RV output index (mL/min/g)	0.50±0.05	0.72±0.08*	0.32±0.02	0.50±0.05*
End systolic elastance (mmHg/mL)	170±30	110±20	300±40	220±20
End diastolic elastance (mmHg/mL)	22±5	13±2	54±11	47±11
Tau indexed (ms/ms)	99±3	112±5*	108±8	105±4

*= p<0.05 vs VEH

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

NRG1 substantially improved cardiac output in the PL group on both time points, without increasing contractility (endsystolic elastance). In contrast, diastolic function showed a trend (p=0.09 at 14 days) to be improved in the NRG1 groups than in the VEH groups (lower enddiastolic elastance indicates less myocardial stiffness). Also, tau (the phase of active ventricular relaxation) was longer in NRG1 than in VEH at 14 days.
Conclusion: In experimental early childhood RV pressure load, NRG1 improves cardiac output independent of contractility, while preserving diastolic function. This challenges the paradigm of contractility-afterload coupling.

O12-5
RNA Expression Profiles and Regulatory Networks in Human Right Ventricular Hypertrophy due to High Pressure Load

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Introduction: Right ventricular hypertrophy (RVH) and remodeling in high pressure afterload, for example in pulmonary arterial hypertension (PAH) or tetralogy of Fallot/pulmonary stensosis (TOF/PS), are associated with alterations in energy metabolism, neurohormonal and epigenetic dysregulation, and a reset of the developmental transcriptional program. We recently identified several interdependent mechanisms such as impaired lipid metabolism, epigenetic miRNA dysregulation, and revival of a fetal gene program, in the SuHx rat model of PAH/RVH/RV failure and in human endstage PAH/RVH/RV failure. However, RNA expression profiling in human non-failing (compensated) RVH has not been performed, and thus RVH-specific regulatory networks are largely unknown.
Methods: We studied intraoperative RV tissue from 19 infants with TOF/PS and RVH (age 2–8 months) and 8 non-RVH age-matched control infants with ventricular septal defects (VSD; 2–12 months). RNA was extracted and sequenced, capturing mRNA, lncRNA, and circRNA (≥10 Gb of cleaned data, 5 million pairs of 100 bp PE reads). The reads were aligned to the GRCh38.p10 human genome reference using STAR, followed by differential expression analysis (EDASeq/DESeq/STARChip).
Results: Using GO-Elite we performed over-representation analysis of the differentially expressed genes in KEGG pathways, cellular biomarkers, and GO-terms. We found differentially expressed genes (RVH vs. no-RVH), significantly overrepresented in pathways related to MAPK signaling, extracellular matrix (ECM)-receptor interaction, focal adhesion and adherens junctions. Additional IPA analysis revealed perturbations in inhibition of matrix metalloproteases, iron homeostasis signaling, tight junction signaling, cardiomyocyte differentiation via BMP receptors, and apelin cardiac fibroblast signaling pathways.
Conclusions: To the best of our knowledge, this is the first unbiased, comprehensive RNA-Seq study of mRNA expression patterns in human RV hypertrophy (here: tetralogy of Fallot). Multiple genes and pathways identified overlap with the mRNA signature we had previously identified in rat and human adult RV failure. Our results advance our current understanding of RV hypertrophy and progressive RV failure, and highlight future therapeutic targets. The upcoming analysis of lncRNA and circRNA expression will allow us to investigate further the related complex transcriptional regulation and RNA biology specific for human RV hypertrophy in tetralogy of Fallot (in the absence of RV failure).

O12-6
Association between genetic variants in the HIF1A-VEGF pathway and left ventricular regional myocardial deformation in patients with Hypertrophic Cardiomyopathy

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Objectives: This study investigated the relationship between left ventricular myocardial deformation and modifier gene risk alleles in HIF1A and VEGF in pediatric patients with hypertrophic cardiomyopathy. Information on genetic etiology of the most common inherited cardiomyopathy related to sudden cardiac death in young people, hypertrophic cardiomyopathy, (HCM) rarely aids in risk stratification and prediction of disease onset, severity or mortality. Little data exist on correlation between genetic modifiers and phenotypic expression of myocardial performance, hampering an individual precision medicine approach.
Methods: In a prospective cohort study, unrelated HCM cases <18 years old were enrolled at the Hospital for Sick Children Toronto (2007-10). Detailed echocardiographic evaluation of LV systolic and diastolic myocardial deformation was assessed using 2-D speckle-tracking echocardiography. From the deformation curves, the LV lateral and septal wall longitudinal peak systolic and diastolic, radial peak systolic and diastolic and circumferential peak systolic and diastolic strain (S) and strain rate (SR) were measured in three lateral, three septal as well as anterior, inferior and posterior segments to reflect multiplane LV myocardial deformation. SNP genotyping for six previously established disease risk alleles in the HIF1A-VEGF pathway was performed in a cohort of pediatric patients with HCM. Findings were correlated with echocardiographic parameters of systolic and diastolic myocardial deformation.
Results: Thirty-two children (6.1±4.5 years; 69% male) with phenotypic and genotypic (59%) HCM were included. Out of six risk alleles tested, one, VEGF1 963 GG, showed a correlation to reduced regional systolic and diastolic LV myocardial deformation. Moreover, LV average and segmental systolic and diastolic strain and strain rate were significantly reduced, as assessed by the standardized difference, in patients harbouring the risk allele.
Conclusions: This is the first study to identify a correlation between a risk allele in the VEGF pathway and regional LV myocardial function; with the VEGF1 963 GG allele associated with reduced LV systolic and diastolic myocardial performance. While studies are needed linking this information to adverse clinical outcomes, this knowledge may help in risk stratification and management of individual patients with HCM.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

O13-1
Comprehensive assessment of outcome in percutaneous device closure of congenital isolated ventricular septal defects in > 400 cases: A single center retrospective database study

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Introduction: Percutaneous device closure of a ventricular septal defect (VSD) is an alternative to surgical treatment in selected cases. Ductal occluders are other alternatives used in our institute to close VSD, which are cost effective and seemed safer. In order to identify suitable cases and reduce failure and complication rates of percutaneous VSD closure, we aimed to 1) study causes of device failure in percutaneous VSD closure and 2) compare outcomes with different VSD types and devices in a high-volume single centre.
Methods: Retrospective data of elective percutaneous VSD closure of isolated congenital VSDs in the 2003-2017 period was analyzed. Outcome was assessed using echocardiography, electrocardiography, and catheterization data. Echocardiography and electrocardiography were performed before procedure, immediately after procedure and during follow up period. Any complications and reinterventions were noted. Logistic regression analyses were used to assess effects of age, VSD type, device type and device size.
Results: During the study period, percutaneous VSD closure was attempted in 412 patients. In n=363 patients VSD closure was successful, in n=30 device implantation failed, and in n=19 the procedure was abandoned because angiographically the VSD seemed unsuitable for device closure. Median(range) age and body surface area were 6.6 years [4.1-10.9] and 0.7 m2 [0.5-1.0] respectively. Device failure was not associated with manufacturer (p=0.09) nor was there a significant difference in failure rate between muscular and ductal devices (p=0.33). Device failure was not associated with age (p=0.08), type of VSD (p=0.5), device type (p=0.2), or device size p=0.1. We observed very low incidence of complete atrioventricular block (0.3%), severe aortic regurgitation (0.3%), and severe tricuspid regurgitation (0.3%).
Conclusion: In our high-volume centre, failure of percutaneous VSD closure occurred in <10% of patients. Because device type is not related to failure rate, it is justified to use the financially beneficial ductal devices in VSD position. Considering the absence of age related risk of device failure or complications, it does not seem necessary to postpone percutaneous VSD closure in symptomatic moderate shunts in younger children.

O13-2
Histopathology of 56 human explanted atrial septal defect (ASD) closure devices

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Objective: To evaluate and characterize tissue reactions within and at the surface of devices for interventional closure of atrial septal defects.
Materials and methods: Explants were processed using a uniform protocol after surgical removal from humans. Devices were fixed in formalin and embedded in methylmethacrylate. Serial sections were obtained by sectioning with a diamond cutter and grinding, thus saving the metal/tissue interface for histological evaluation. Immunohistochemical stainings were performed using conventional protocols.
Results: 56 devices were analysed (Amplatzer n = 28, Cardioseal/Starflex n= 9, Biostar n= 5, Helex n= 4, others n= 10). Implants had been in the human body for 1 day to 15 years (mean 3.2 years). Main reason for explantation had been residual shunting in 19, thrombi and/or emboli in 10, heart surgery otherwise indicated in 8, device dislocation in 7, deformity of the device in 5, damage to the right atrial wall in 3 and other reasons in 4 patients, respectively. Endothelialisation and cellular organisation of tissue within the devices was present in all specimen with implantation times > 6 months. Lymphocytic infiltrations and local foreign body reaction related to textile components were found in almost all explants. Calcifications and partial corrosion or even loss of "permanent" materials (metal wires, ivalon foam) were seen in explants with implantation time > 7 years.
Conclusion: This is the largest cohort on ASD occlusion devices with complete histology work-up after surgical explantation. We demonstrate timely endothelialisation and tissue organisation, a typical pattern of chronically persisting inflammation. Calcifications and material alterations were seen in long-term explants. We conclude that patients with ASD occlusion devices should be followed life-long for detection of potential implant-related complications. Our findings may be relevant for development of new devices.

O13-3
The size is not the matter: Percutaneous pulmonary valve implantation in extreme small conduits

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Background: Guidelines allow percutaneous pulmonary valve implantation (PPVI) with the Melody valve, in dysfunctional right ventricular outflow tract (RVOT) conduits >16 mm in diameter at the time of implant. It is not clear that small original conduit diameter should be an a priori exclusion criterion for PPVI. Limited data are available regarding the ability to enlarge conduits substantially beyond the original diameter or with significant conduit wall injury.
Methods: All patients with an original (implanted) expandable RVOT conduit diameter <16 mm who underwent percutaneous catheterization for intended PPVI at our institution from March 2007 to November 2018 were analyzed for this study.
Results: A total of 31 patients met inclusion criteria and 20 were finally included, (11 of 31(35%) patients had a conduit that was larger than the reported implant diameter (1 pulmonary homograft, 10 Contegra) and were excluded). Median age and weight of the 20 patients was 9,9 (3,4-17) years and 33 (13-81) kg. The median original conduit diameter was 13 (11-15) mm, and the median narrowest conduit diameter was 11,7 (7-14,9) mm. Conduits were enlarged to a median diameter of 20,4 mm (42% larger than the implanted diameter). Largest balloon to measured conduit diameter ratio was 166% (129-257). There was significant hemodynamic improvement post-implant, median RV/Ao pressure ratio pre-procedure and post-procedure was 65% (37-104) and 42% (24-88), and no significant residual peak RVOT pressure gradient and pulmonary regurgitation. During a median follow-up of 2.7 years, freedom from RVOT reintervention was 100% and 95% at 2 and 4 years, respectively, and there were no deaths and 2 cases of endocarditis.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusion: In our experience, PPVI with the Melody valve into small conduits (<16mm), was feasible and safe, with favorable outcomes, that did not appear to differ dramatically from published series in larger conduits and valves. We achieved a median diameter 42% larger than the nominal conduit implanted (29% previously reported in a multicentric study) without significant conduit wall injury.

O13-4
Percutaneous closure of patent ductus arteriosus in premature infants: a French national survey

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Background: Transcatheter closure of PDA in premature infants has been shown to be feasible in small series. Outcomes in large series is currently lacking.
Material: All premature infants (< 36 weeks GA) who underwent transcatheter PDA closure were included in a multicenter French national survey. Demographic data (gestational age (GA), birth weight (BW)) as well as procedural data (weight (PW), age at procedure (AP), procedural success, fluoroscopy time and type of device) were collected. Outcomes and procedural complications were reviewed.
Results: Between September 2013 and June 2017, 102 patients were included. In 71 cases, PDA pharmacological closure had previously been attempted. Mean GA was 27 +/-2.9 weeks. Mean BW and PW were 1040+/-715 g and 1543+/-698 g, respectively. Mean AP was 39+/-26 days. Number of premature infants below 1 kg, between 1 and 2kg and above 2 kg was 21, 59 and 22, respectively. AP was significantly lower in patients with lower weight. Mean fluoroscopic time was 6.5 minutes. Success rate was 99%. One PDA was too large to be closed with unstable device. Device or procedure related complications were reported in 9 patients (8.9%) including three LPA stenoses (requiring surgery in 2 and balloon dilatation in one), two neo-coarctations (one requiring subsequent surgery), and 3 tricuspid valve regurgitations at follow-up. Seven deaths were reported, none being related to the procedure. Mean follow-up was 39.75 +/-13.1 months.
Conclusion: In this large series of premature infants, efficacy of transcatheter PDA closure has been good with an acceptable rate of procedural complications.

O13-5
Outcomes after percutaneous vs. surgical pulmonary valve implantation – up to 12 years follow up of 481 patients from a single center

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Introduction: Percutaneous pulmonary valve implantation (PPVI) was introduced initially to expand the lifetime of a surgically implanted pulmonary valve by at least two years. Studies comparing PPVI and surgical pulmonary valve replacement (SPVR) in terms of survival and reinterventions at the long-term are missing.
Methods: The clinical information of all patients who were treated by PPVI and SPVR in our center between 12.2006 and 12.2018 was prospectively enrolled into a database. SPVR was only done in patients who were considered to be unsuitable for PPVI. We evaluated the patients' long term follow up data in means of survival and further valve replacement.
Results: The study comprised 481 patients. Percutanous pulmonary valves were implanted in 265 patients and 216 patients were treated with SPVR. In the PPVI cohort, 242 (91%) recieved the Melody valve and 23 patients (9%) – the Sapien valve. The patients with SPVR were treated with biological valves: Homograft (n=139; 64%), Hancock (n=57; 26%), Contegra (n=13; 6%) and others (n=7; 3%). There were no significant differences in the age and weight of the patients from both groups. Median follow up was 4 years (range 6m – 12years). Seventeen patients died (3.5%), seven after PPVI, ten after SPVR (p=0.24). Estimated survival at 10 years after PPVI was 95%, which was comparable to that after SPVR – 94%, p=0.43. Estimated survival with the originally implanted valve at 10 years was 86% in the PPVI group vs. 72% in the SPVR group, (p=0.27). In the surviving patients, freedom from reinterventions on the valve because of infective endocarditis at 10 years was 95.7% for the PPVI cohort vs. 97.3% for the SPVR group, p=0.174.
Conclusion: Long-term survival and freedom from reinterventions because of degeneration or endocarditis after PPVI is comparable to that after SPVR. Percutaneous pulmonary valves show excellent longevity which extends to more than ten years. Thus, due to its less invasive approach, PPVI is the method of first choice whenever possible to treat pulmonary valve or conduit failure.

O13-6
Sparing the pulmonary valve during RVOT stenting in Fallot – is it worth the effort?

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Introduction: Stenting of the RVOT is a safe and effective technique in the initial palliation of selected patients with Fallot type lesions. Stenting of the RVOT increases pulsatile forward flow of systemic venous blood to the PAs. This results in a greater rise in systemic oxygen saturations and promotes better pulmonary arterial growth compared to BT Shunt palliation.
There is debate as to whether the pulmonary valve should be crossed or spared during RVOT stenting. Conceptually, not crossing the PV should have many advantages: potential for the pulmonary valve to grow, avoidance of free regurgitation and potential of later repair without transannular patch.
Objective: To assess the outcomes of stenting the right ventricular outflow tract (RVOT) in patients with Tetralogy of Fallot whilst sparing the pulmonary valve.
Methods: Retrospective, non-randomised, single centre review of patients with Tetralogy of Fallot/AVSD Fallot who underwent RVOT stenting between 2008-2017 and came forward for delayed complete repair. Pulmonary valve growth was assessed by serial echocardiography.
Results: 58 patients were studied. Stents were placed crossing the valve in 28 patients and sparing the valve in 30 patients (52%).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

There was significant growth of the PV after valve sparing RVOT stent [$p < 0.01$; two tailed t-test].
Valve preserving Fallot repair was achieved in 4 cases (13%) of cases after valve sparing stent. There was no difference in the rate of transannular repair between the 2 groups. There was a lower need for conduit repair in the valve sparing stent group (23% vs 40%) [$p < 0.03$]
Conclusions: Initial palliation of Fallot is required in about 20% of cases. Stenting the RVOT compares favourably to BT shunt. Stenting the RVOT in Fallot lesions without crossing the pulmonary valve promotes growth of the PV annulus and thereby facilitating valve sparing corrective surgery at a later stage. This approach should be favoured in cases with hypoplastic pulmonary arteries, anomalous coronary arteries or those with associated cardiac lesions or syndromes.

O14-1
In-utero aortic valvuloplasty in fetuses with critical aortic stenosis and evolving hypoplastic left heart syndrome: effects on the fetal heart and on outcome. A single center analysis of 80 patients

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Objectives: to assess effects of fetal aortic valvuloplasty (FAV)in patients with critical aortic stenosis (CAS) and evolving hypoplastic left heart syndrome (eHLHS) on left ventricular (LV) dimensions, function and outcome and to look for predictors of biventricular (BV)outcome.
Methods: Echocardiograms of all fetuses who underwent FAV in our center since 2001 were analyzed retrospectively for ventricular and valvular dimensions, mitral regurgitation (MR) velocity, LV filling time and outcome (BV vs. univentricular (UV)).
Results: 95 FAV were performed in 80 patients (success-rate: 86.3%). Median GA: 26+3 weeks (21+3 to 33+1). 62 successfully treated patients were live-born and 74.6% of neonates were initially treated towards a BV circulation. After 28 days 42/58 (72.4%) and at final follow up (median 2 years (32 days to 13 years) 35/51 (68.6%) were alive with a BV circulation without elevated pulmonary artery pressure. BV outcome was significantly better when compared to a natural history cohort with similar inclusion criteria (10/35, 28.6%; $P = 0.0004$).
When the preintervention data was compared between the BV and the other groups (BV vs. UV/dead with BV) significant differences could be found in these parameters:
Successful intervention led to an immediate increase in RV/LV ratio in both groups ($P = 0.0001$ vs. $P = 0.05$) due to reduced LV lengths and an increased LV inflow time ($P \leq 0.0000001$ vs. $P = 0.004$).
Conclusions: Successful FAV in fetuses with CAS and eHLHS improved morphologic and functional LV-parameters and BV outcome when compared to published natural history data. Existing criteria for intervention have to be further refined for a better patient selection and prospective studies are warranted.

	BV (n=25)	UV + others (n=17)	P
RV/LV	1.073 (0.907-1.129)	1.173 (1.06-1.485)	≤ 0.00001
TV/MV	1.239 (1.089-1.453)	1.375 (1.237-1.594)	= 0.00005
LV inflow time	0.29 (0.12-0.49)	0.19 (0.12-0.43)	= 0.035
MR velocity	3.59 (2.00-5.07)	2.70 (1.55-4.64)	= 0.007

O14-2
Lessons from prenatal diagnosis and in utero transfer of fetuses with transposition of the great arteries

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Background: Prenatal diagnosis of transposition of the great arteries (TGA) reduces neonatal mortality and morbidity. The mortality of prenatally diagnosed TGA remains scarcely described.
Objective: To review the experience of prenatal diagnosis and in utero transfer of fetuses with TGA to assess the limits of this program and to describe outcomes of prenatally diagnosed TGA.
Methods: From 1999 to 2017, we reviewed all cases of prenatally diagnosed TGA born at our institution. Files were analyzed for discordances between prenatal and postnatal diagnosis, status at birth, neonatal management, and outcomes.
Results: During the study period, 748 fetuses with prenatal diagnosis of TGA (470 simple, 278 complex) were delivered at our institution and 333 neonates with post-natal diagnosis were referred after having been delivered elsewhere. Discordance between pre- and postnatal diagnosis was noted in 14.8% with two third having consequence on surgical treatment. Median term was 39 WG. The proportion of premature delivery (<37WG) was 5.3%, the majority being late-preterm (>34WG). 100 neonates were small for gestational age (<10th percentile); 10 from twin pregnancies and 10 with very low birth weight (<5th percentile). 14% were intubated in the delivery room and 1.7% required resuscitation maneuvers. 64% had a Rashkind procedure that was done in 15% of cases in the delivery room. 55% received PGE1 infusion and 38% had a Rashkind and PGE1 infusion. There were 9 deaths before surgery (1.2%) with two not related to the TGA (1 foeto-maternal hemorrhage and 1 polymalformation syndrome). A fourth of the cohort experienced neonatal complications including mainly respiratory distress requiring ventilation, infection, necrotizing enterocolitis, pulmonary hypertension and iatrogenic events. Surgical mortality was 1.8% (4 simple TGA and 10 complex). Seven additional deaths occurred during the first year follow-up, 6 being related to late cardiac complications. Overall survival at one year was 96%.
Conclusion: Prenatal diagnosis and in utero transfer of fetuses with TGA does not eliminate the risk of pre-operative mortality. The proportion of immediate distress is high and the need for intensive care in the delivery room is significant. Finally, preoperative mortality accounts for a third of the one-year mortality in this population.

O14-3
Postnatal outcome in antenatally diagnosed atrioventricular and ventriculoarterial discordance

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Introduction: There are few published data describing the outcome of fetuses with discordant atrioventricular and ventriculoarterial connections (DAVVAC). We aimed to describe survival and postnatal interventions in a large cohort of antenatally diagnosed DAVVAC, and identify antenatal predictors of survival.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Methods: Fetuses with at least one discordant atrioventricular and one discordant ventriculoarterial connection were identified from the databases of two centres of fetal cardiology for the period 01/01/1989 – 01/06/2018, and antenatal and postnatal data were collected. Data were analysed using Kaplan Meier analysis with the Mantel-Cox test.
Results: 98 fetuses were identified. 39 pregnancies were terminated, there were no intrauterine deaths, and 16 were lost to follow up. Postnatal data were available for 43 patients, with a median follow-up of 9.5 years and maximum of 23 years. There were 8 deaths, 5 during first year of life, and all before the age of 4 years. The cohort was divided into 5 groups: 15 patients with no additional structural cardiac lesions (isolated), 17 with a ventricular septal defect (VSD) only, 21 with pulmonary stenosis +/- VSD, 8 with Ebstein's anomaly of the tricuspid valve +/- VSD, and 37 patients with other cardiac abnormalities. The best survival was seen in the pulmonary stenosis group, with no deaths. One death was seen in the isolated group, due to complete heart block that developed postnatally. The worst survival was seen in the Ebstein's group which was significantly worse than the rest of the cohort as a whole (p=0.02). 6 patients underwent permanent pacemaker implantation, and 23 underwent at least one other cardiac surgical procedure. The presence of antenatal tricuspid valve regurgitation was associated with worse postnatal survival (p=0.002). Antenatal complete heart block was seen in 4 patients, and the survival of this group was worse (although this did not reach statistical significance).
Conclusions: This is the largest cohort described of antenatally diagnosed DAVVAC. The medium-term outcome of isolated DAVVAC and DAVVAC with pulmonary stenosis is very good, and the outcome of DAVVAC with Ebstein's anomaly is poor. Antenatal tricuspid valve regurgitation is associated with a worse postnatal outcome.

O14-4
When is a fetal persistant left superior vena cava truly isolated? A retrospective cohort of 230 cases

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Basis: Isolated persistent left superior vena cava (PLSVC) is not associated with any cardiac, extra-cardiac or genetic anomaly. The definition of isolated PLSVC varies as some anomalies can be difficult to detect antenatally. Even if not clinically relevant for the patient some of them may be related to a genetic anomaly.
Objectives: To assess the frequency of genetic, cardiac or extra cardiac anomalies in fetuses with isolated versus associated PLSVC.
Methods: Retrospective cohort study including all fetuses diagnosed with a PLSVC between 2010 and 2017. PLSVC was categorised as isolated or associated according to the ante natal diagnosis of associated congenital heart defects, abnormal venous or arterial connections, and/or extracardiac anomalies.
Results: Among 230 fetuses diagnosed with PLSVC, 40 cases (17.4%) were strictly isolated and no syndromic/genetic anomaly or coarctation of the aorta was diagnosed. In the remaining 190 fetuses with PLSVC and associated prenatal ultrasound features, 65 (34%) fetuses had a genetic anomaly: 23 aneuploidies (10 trisomies 21, 7 trisomies 18, 2 trisomies 13, 4 Turner syndrome), 15 pathogenic micro-deletions/duplications (including 3 deletions 22q11.2) and 5 variants of unknown significance. In particular, PLSVC associated with abnormal venous or arterial connections (aberrant sub clavian artery, abnormal ductus venosus) presented a 22.2% rate of genetic anomalies. Small ventricular septal defect, single umbilical artery or hypoplastic aortic isthmus were not associated with higher rate of genetic anomaly. The number of aortic coarctations was low overall (7/230). However, among fetuses without evidence of a major congenital heart defect or extracardiac anomalies, z score of the aortic isthmus diameter inferior to -2 was significantly associated with the postnatal diagnosis of isthmus anomalies (10/19 versus 2/52, p<0.001).
Conclusions: Fetuses with strictly isolated PLSVC did not show any genetic or cardiac anomaly. A careful cardiac and morphologic ultrasound examination is paramount as minor variants of the venous or arterial system may increase the index of suspicion for genetic abnormalities and invasive prenatal diagnosis for array-comparative genomic hybridization should then be offered.

O14-5
Does 3D/4D spatiotemporal image correlation (STIC) echocardiography improve diagnostic accuracy and predicting surgical approach in fetuses with double outlet right ventricle?

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Background:: DORV is a complex form congenital heart disease which is heterogeneous with respect to location of interventricular communication(IC) and the relationship of the great arteries.
Aim: To analyze the incremental benefit of 3D/4D STIC fetal echocardiography with offline processing in improving diagnostic accuracy and predict surgical approach in fetuses with DORV.
Methods: Our database was reviewed retrospectively from January 2008 to October 2018.Cases which underwent termination of pregnancy and those without postnatal data were excluded. 3D/4D STIC fetal echocardiography with offline analysis was included in the protocol after October 2015. Fetal diagnosis was compared with post-natal echocardiography with respect to situs, IC, great artery relationship to IC, outflow tract anatomy & presence/absence of two ventricles. The incremental benefit of 3D/4D STIC fetal echocardiography on diagnostic accuracy (> 4/5 parameters) when compared to conventional fetal echocardiography was studied. The accuracy of fetal echocardiography in predicting the surgical approach was analyzed.
Results: Of the total 138 cases with diagnosis of DORV in fetal echocardiography during the study period, 61 (44%) were delivered in our centre and had post-natal evaluation. The mean gestational age was 30.1 + 5.7 weeks. Of these, 35 had only conventional fetal echocardiography while 26 had in addition 3D/4D STIC imaging also. Ten patients (7 in conventional and 3 in 3D/4DSTIC) were deemed not suitable for biventricular repair and were excluded from further analysis. The comparison of diagnostic accuracy of conventional vs. 3D/4D STIC for various anatomic components is as follows:
The overall diagnostic accuracy was superior with 3D/4D STIC compared to conventional fetal echocardiography (88% vs. 60%; p = 0.020). Accurate prediction of single staged biventricular repair was made in 60% of cases with conventional echocardiography and all cases (100%) with 3D/4D STIC imaging (p=0.024).

Variable	Conventional	3D/4D STIC
Situs	91%	95.6%
VSD location	65%	87%
Great artery relation	56%	87%
Outflow obstruction	87%	100%
Biventricular anatomy	65.7%	88%

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusion: Addition of 3D/4D STIC fetal echocardiography to the conventional imaging provides incremental diagnostic accuracy in fetuses with Double Outlet right ventricle, thereby aiding in counseling and planning surgical approach.

O14-6
Fetal mean pulmonary artery pressure falls and pulmonary vascular maturation improves after reversal of ductal constriction: a Doppler echocardiographic study

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Introduction: Constriction of ductus arteriosus is prevalent in third trimester fetuses, as a result of maternal utilization of anti-inflammatory pharmacological or dietary prostaglandin inhibitors, with habitual reversal after suspension of the causal agent. Improvement of pulmonary hypertension and pulmonary vascular maturation after this reversal has not been previously demonstrated in human fetuses. This study was designed to test the hypothesis that estimated mean pulmonary artery pressure (MPAP) decreases and pulmonary vascular maturation assessed by acceleration time/ejection time ratio (AT/ET) increases after reversal of ductal constriction and that these effects are independent of gestational age evolution.
Methods: This is a prospective observational study, comparing Doppler echocardiographic ductal flow dynamics parameters, MPAP and AT/ET ratio in 53 third trimester fetuses at the moment of ductal constriction diagnosis and after 2 weeks of discontinuation of prostaglandin inhibitors to a control group of normal fetuses from local nomograms. MPAP was estimated by Dabestani equation and vascular maturity by AT/ET ratio, according to reported validations. Statistical analysis utilized t test for comparison of the variables at diagnosis and after reversal of ductal constriction. Variations of MPAP and AT/ET ratio at these 2 moments were compared to the normal expected variations at the same gestational period.
Results: Normalization of mean systolic and diastolic ductal velocities (1.85±0.27 to 1.38±0.39 m/s, $p<0.0001$, and 0.43±0.10 to 0.21±0.06 m/s, $p<0.0001$, respectively) and of pulsatility index (1.98 ± 0.20 to 2.60 ± 0.30, $p<0.0001$) was demonstrated after 2 weeks. In this period, mean MPAP decreased (65.0±7.2 to 53.4±6.9 mmHg, $p< 0.0001$), and AT/ET ratio increased (0.19 ±0.06 to 0.33 ±0.07, $p< 0.0001$). Variation of mean MPAP was -12,5 ±7,5 mmHg, $p<0.001$ (normal variation = -1.3 ± 0.19 mmHg,[9.6 times more], $p<0.001$),and variation of pulmonary AT/ET ratio was +2.12 ±0.48, $p<0.001$ (variation of AT/ET in normal fetuses = + 0.13 ±0.08[16 times more], $p<0.001$).
Conclusion: This study shows for the first time that resolution of fetal ductal constriction is followed by fall in the MPAP and increase in pulmonary vascular maturity, at a significant higher degree than the observed in normal fetuses in the same gestational age variation period.

O15-1
Percutaneous Closure of Atrial Septal Defects in Elderly patients: the experience of a high-volume referral tertiary center

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Objectives: Ostium Secundum Atrial Septal Defects (ASDs) are a common congenital heart disease. An early diagnosis allows to close them before to develop a pulmonary hypertension. In some cases, these defects are not diagnosed in childhood, but when symptoms and signs compare and pulmonary arterial pressure increases. The elderly patients (>60 years) are a complex subgroup to treat with several complications and hemodynamic limitations.
Methods: From March 2000 to November 2018, 1084 patients (pts) with ASD underwent closure at our institution. 65 were elderly patients (≥ 60 years). Mean age and weight were 65,07±3,82 years (range 60-75) and 71,27±12,35 kg (range 45-105), respectively. Right heart catheterization was performed to evaluate pulmonary pressures, pulmonary vascular resistances and to address the closure.
Results: Among these elderly patients: 28 (43%) had high mean pulmonary arterial pressure (mPAP≥25mmHg). At ultivariate analysis, the mean pulmonary pressure was not influenced by the age ($p=0.8$) but was mainly correlated with ASD dimension ($p<0.01$). At admission, 27 patients (41,5%) showed atrial flutter or fibrillation. Supraventricular arrhythmias were mainly associated with elevated mean pulmonary pressure ($p<0.01$), whereas the age didn't influence them. One patient with high pulmonary pressure (mPAP=44mmHg) and high arteriolar pulmonary vascular resistances (PVR=3,15WU) underwent sildenafil therapy for 3 months with a significant improvement of hemodynamic data (mPAP=25mmHg and PVR=0,65WU) and consequent ASD closure. In 3 cases ASD wasn't closed because of pulmonary hypertension with high arteriolar pulmonary vascular resistances (PVR>3WU). Percutaneous closure was effectiveness in all resting patients and no major complications were recorded. At mean follow-up of 4,3 years, no mortality or morbidity related to procedure arose and no patients developed new onset arrhythmias.
Conclusion: ASD closure in elderly patient is often challenging for interventional cardiology because of the high pulmonary pressures. Pulmonary pressure is related to ASD dimension (not to the age), and the onset of atrial flutter or fibrillation is influenced by high pulmonary pressure. The right heart catheterization is helpful to guide decision making of interventional cardiology. No mortality or morbidity related to procedure were showed during follow-up.

O15-2
Early outcomes of percutaneous pulmonary valve implantation using the Edwards Sapien 3 transcatheter heart valve system – German experience

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Background: Percutaneous pulmonary valve implantation (PPVI) is an increasingly practiced treatment option for patients with right ventricular outflow tract dysfunction. After encouraging results with the Edwards Sapien and XT valves in the pulmonary position, Edwards' latest modification, the Sapien 3 valve is available for clinical PPVI trials.
Objectives: This study aimed to review procedural data and early outcomes for the Sapien 3 valves (Edwards Lifesciences, Irvine, California) for PPVI.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Methods: We performed a multicenter, retrospective, observational registry analysis of patients who underwent PPVI with the Edwards Sapien 3 transcatheter heart valve between 2015 and 2017 in 5 centers in Germany.
Results: 46 patients could be enrolled (mean weight 56,8 ±26,9 kg, min. 11,8, max. 114 kg). The majority had tetralogy of Fallot as underlying diagnosis (48%), and a Contegra conduit as the most common RVOT configuration pre PPVI (34,8%). However, pulmonary insufficiency or both, insufficiency ≥ moderate and stenosis ≥ 20 mmHg were the leading indications for PPVI (78,3%). Most procedures were 2-stage procedures (82,6%) with 100% pre-stenting. Valve sizes were 20 mm (n = 1), 23 mm (n = 15), 26 mm (n = 18), 29 mm (n = 12). Procedural success rate was high (95,6%) with a low frequency of periprocedural complications (4,3%): In 2 patients surgical pulmonary valve implantation had to be performed after balloon rupture during (one-stage) PPVI procedure.
Follow-up data was available up to 24 month post PPVI. NYHA class improved in all patients (93,3% were at NYHA I). The rate of patients with moderate/severe pulmonary regurgitation decreased from 74% at baseline to 0% after PPVI, and the calculated peak systolic gradient for all patients decreased from 24.2 (SD±20.9) mmHg to 7,1 mmHg (SD±5,0). There were no episodes of endocarditis, no thromboses and no stent fractures documented.
Conclusions: The Edwards Sapien 3 valve is a viable option for PPVI in patients with conduits, native pulmonary valves or transannular patches. Continued data collection is necessary to verify long-term results.

O15-3
Building contingency: emergency cardiac surgery following interventional treatment of congenital heart disease

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Introduction and and objectives: Interventional catheter based management has transformed management of congenital heart but carries the risk of significant complications requiring surgical intervention. Our objectives were to review our experience of emergency cardiac surgery following interventional treatment of congenital heart disease (CHD) and identify common themes.
Methods: A retrospective review of all unit data submitted to the externally validated national congenital cardiac audit database between April 2001 and September 2018 was performed to identify patients who underwent unplanned surgery within 24 hours of catheter intervention. The data was cleaned to identify only those patients who underwent unplanned surgery because of (i) a complication or (ii) unsuccessful intervention but no complication.
Results: There were 3824 interventional catheter procedures of which 20 required unplanned surgery within 24 hours. The median age was 0.27 years (range 0-47.6) and median weight 4.3 kg (range 2.5-56). There were 14 in group (i) and 6 in group (ii). In group (i), 11 underwent immediate surgery and 3 had surgery the next day. In group (ii), 3 underwent immediate surgery and 3 had surgery the next day. Of group (i), 8 involved retrieval of a migrated or misplaced device (pulmonary outflow stent: n=3, percutaneous valve: n=2, duct, atrial septal or pulmonary artery occluder: n=3), 3 had tamponade after attempted radiofrequency perforation of the pulmonary valve, 1 patient had acute conduit obstruction post angioplasty, 1 had ASD device removal for tamponade and 1 had tricuspid valve repair after pulmonary valve dilatation. Of group (ii), 3 remained desaturated after pulmonary valve dilatation, 1 had a failed balloon septostomy, 1 had a one poor response to aortic valve dilatation and 1 had failed dilatation of aortic recoarctation. 2 patients died within 30 days, 1 from group (i) following migrated percutaneous valve and 1 from group (ii) following a Norwood procedure.
Conclusions: The need for unplanned cardiac surgery after congenital catheter intervention is rare, however a significant proportion require emergency surgery underlying the need for on-site cardiac surgical support. Common themes in our population were younger patient age and procedures to the right ventricular outflow tract.

O15-4
Medium- and long-term follow-up of transcatheter closure of ruptured sinus of Valsalva aneurysm

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Introduction: We aim to evaluate medium- and long-term outcomes of transcatheter closure (TC) of ruptured sinus of Valsalva aneurysm (RSVA), which is a rare and mostly congenital heart disease.
Methods: Retrospective analysis included 23 patients (pts; 14 males) aged 15-79 years (y; 39.9±18.5) selected for TC of RSVA between 2007 and 2017 in two tertiary centres. 15 pts were in NYHA class III or IV before TC, 5 pts had acquired RSVA after previous cardiac surgery. Echocardiography revealed rupture of right/noncoronary sinus to right atrium in majority of pts (17/23). Qp/Qs ranged from 1.4 to 3.7 (median 2.2). Defect's aortic orifice diameter was 9.5 ± 3.3 mm (4-16). Fluoroscopy time was 18.0 ± 11.0 minutes (5-48). We applied 22 duct, 3 muscular and 1 atrial septal Amplatzer or Amplatzer-like occluders by anterograde venous approach after arterio-venous loop creation in all but 1 pt. Mean follow-up conducted in outpatient clinic was 5.5 ± 3.5 y (range 1-11).
Results: The procedure was successful in 19/23 pts (82.6%). Four procedures were abandoned and the devices were percutaneously retrieved because of coronary artery compression (1 pt), transient increase of aortic regurgitation (AR; 1 pt) and embolization (2 pts). New onset of significant AR was noted in the one of the latter pts after device removal. NYHA class has improved in all treated pts but 2, in whom remained stable. Three pts needed percutaneous reintervention in follow-up because of significant residual shunt in 1 and late recurrent RSVA in 2 pts. Follow-up of remaining pts was uneventful. Neither erosion, embolization, new AR nor death were observed.
Conclusions: Percutaneous closure of RSVA is a safe and effective method of treatment with good clinical outcome. However, although not described previously, recurrent shunts after TC of RSVA are possible.

O15-5
Stent Implantation for Aortic Coarctation in Children < 6 years: Initial and 8 - years Results

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Introduction: Although stenting has been used as a treatment option for CoA at increasingly younger ages, there is limited information on the long-term follow-up of stent implantation for native CoA in small pediatric patients. This study reports initial and 10-years results following stent implantation for coarctation of the aorta (CoA) in children less than 5-years of age.
Methods: Sixty-three patients with native CoA (NaCoA) (median age 3 years, range 1–5.5 years) underwent stent implantation (SI) using PG2910B and ev3 stents. Bench testing of the stents was performed to determine the smallest sheath diameter that is required for their use. Patients with hypoplasia of the proximal aortic isthmus or transverse aortic arch were excluded from the study.
Results: The stents were crimpted on a 7-10 mm balloon and implanted through a 6–8 sheath. The stents were further dilated to a larger diameter using 10-14 mm balloons that were introduced through a 7 - 8F sheath. 21 (31%) patients with an arm/leg pressure gradient ≥ 20 mm Hg underwent successful stent re-dilation 4 to 8 years after the initial procedure for a relative to growth stenosis. Immediately after stenting the peak systolic pressure gradient fell from 68±16 mmHg to 8±5 mmHg, while CoA diameter increased from 5 ± 3 mm to 16. ± 3 mm. Peak systolic pressure gradient was reduced from 20 2 mm Hg (range 18 to 23 mm Hg) to 5 ± 2 mmHg (range 0 to 7 mmHg) after re-dilation. There were no major procedural complications. Late aneurysm formation and stent fracture that required a new stent implantation were observed in 2 and 3 patients, respectively. At the end of follow-up no cases of recoarctation were identified on angiography, or MSCT. Fifty – eight (92%) of the patients were normotensive at the end of follow – up period.
Conclusions: Stent implantation is an effective and safe alternative to conventional surgical management for the treatment of selected pediatric patients with CoA.

O15-6
Tips and pitfalls in transplant surgery after Fontan
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Introduction: Heart transplantation after Fontan completion poses a unique surgical challenge. Seventeen patients are presented, stressing the technical hints performed in the five anastomoses to match the graft in the recipient.
Methods: Data are collected from fifteen Fontan and two takedown patients along 6 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 13 patients, whereas atriopulmonary connection was found in three and lateral tunnel in one. Five patients developed protein losing enteropathy. Thirteen stents have been previously deployed in left pulmonary artery (8), inferior vena cava (3), superior vena cava (1) 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 eight times (hemi-arch repair), superior vena cava five times (one case with double superior vena cavae), pulmonary branches thirteen times (hilum to hilum plasty with donor's aorta/pericardium patch after thorough stent removal) and inferior vena cava twelve times (conduit sleeve anastomoses). Follow-up was complete for a median of 45 months (range 2-70). Two patients died. ECMO was needed in six 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 four children. Interestingly, one 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 dysfunction was needed in one third of cases. Overall results can match other transplants cohorts.

MP1-1
Transcatheter Closure of Moderate to Large Congenital Arteriovenous Fistula Using the Amplatzer Duct Occluders I and II
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Introduction: Transcatheter closure of congenital coronary arteriovenous fistula (CAVF) using coils and occluding devices has become an accepted alternative to surgical closure. The aim of this study was to present data from 33 children with moderate to large CAVF who underwent catheter closure using the Amplatzer duct occluders I and II (ADOI ADO II).
Methods: The median age of the patients was 3 years (range 2- 12 years). The anatomy and size of CAFF were defined by retrograde aortography and/or selective injection of contrast media in the proximal part of fistula. The devices were deployed from the femoral vein with the formation of an arteriovenous loop. The procedure was guided by hand injection proximal coronary arteriography.
Results: The fistula originated from the left coronary artery and the right coronary artery in 21 (64%) and 12 (26%) patients, respectively. The drainage sites were: the right atrium in 18 (54%) patients, the coronary sinus in 8 (24%) patients, and the right ventricle in 7 (22%) patients, respectively. The mean diameter of the CAVF was 5.2 ± 2.3 mm (range 3 – 10 mm). The mean device diameter was 5 ± 2 mm (range 6 – 10mm) and 4.2 ± 1.5 (range 3 – 6 mm) for the ADO I and ADO II, respectively. A 7-8F delivery sheath (DS) was used in 12 (ADO I) and a 4 - 5F in 21 patients, respectively. The occluders were implanted permanently in all patients. Complete occlusion after the procedure was observed in 32/33 (97%) of the patients. Echocardiographic closure was demonstrated in all patients at the 3 – month follow – up. Four loses of the arterial pulses that were restored with intravenous infusion of heparin and rtPA (2 patients) the only complications of the procedure.
Conclusions: The ADO occluders due to their safety, high complete closure rate, easiness of loading and implantation are the devices of choice for closure of moderate to large CAVF. When compared to ADOI the ADO due to its low profile can be implanted through a smaller DS facilitating closure in pediatric patients and tortuous CAVF.

MP1-2
Transcatheter Occlusion of PDA in Extremely Premature Infants Weighting Less Than 1800 g: A Single-Center Experience
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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Introduction: Patent ductus arteriosus (PDA) in premature infants continues to be a significant clinical problem contributing importantly to both morbidity and mortality. Surgical ligation and medical therapy both have their drawbacks. The aim of our study is to describe a single center experience with a new endovascular occlusion device Medtronic Micro Vascular Plug (MVP) used for transcatheter PDA closure in extremely preterm infants.
Methods: Between January 2018 and November 2018, 54 percutaneous PDA closures were done in our Center. Sixteen infants (30%) who weighed less than 1800 g were included in this retrospective study. Procedural details, complications, and short-term outcomes were recorded.
Results: Sixteen premature infants underwent attempted transcatheter PDA closure using the Medtronic MVP. The gestational age and birth weight were 25.0+1.4 (range 24-29) weeks and 768+251 g (range 440-1580), respectively. The mean weight and age at the time of the procedure were 1.32+0.27 kg (range 920-1800) and 46.8+14.3 days (range 24-68), respectively. The mean PDA diameter was 3.2+0.9 mm. All ducts were tubular in nature. All devices were deployed via a 4F Glide catheter in prograde fashion without arterial access. Fluoroscopy and echocardiography were utilized for the procedure guiding placement of the PDA device. Mean fluoroscopy time and radiation dose were 7.5+3.9 min and 15+8.6 mGy, respectively. Initially heparin was given in 3 patients due to decreased pulse; currently heparin is given for all patients (10 units/kg/h for 24 hours) prophylactically. Complete closure was achieved in all infants with no procedural complications (including vocal cord dysfunction), pulmonary artery or aortic obstruction or death. There were no additional complications related to the procedure or noted during short-term follow-up (mean, 130+96 days).
Conclusions: This preliminary study demonstrates that transcatheter PDA closure can be successfully performed in extremely preterm infants weighting less than 1800 g using the Medtronic MVP with a high success rate and a low incidence of complications. Trascatheter PDA closure may be an excellent alternative to surgery.

MP1-3
Mid and long-term follow up after stent implantation for aortic coarctation.

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Introduction: Stent implantation to treat aortic coarctation is nowadays a usual technique in adolescents and adults. There is lack of evidence among the mid and long term results of this technique.
Patients and methods: We performed a retrospective study of the follow-up of the patients with native aortic coarctation and recoarctation treated by stent implantation in our hospital.

Age	16.5 years (IQR 13.58- 21.91)
Weight	58 Kg (SD 18.1)
Time since surgery	15.7 years (SD 7.45)
Type of stent implanted	CP (42.9%)
	CP-covered (17.6%)
	EV3 (14.3%)
	Palmaz (9.9%)
	Other (15.3%)
Follow-up	6.6 years (SD 4.34) (maximum 18.4 years)

Results: Between April 1997 and June 2018, 95 patients were treated with stent implantation (69 recoarctations and 26 native). We analyzed the mid and long term outcome of the 68 patients with a follow-up longer than 3 years. The follow up visits included EKG and echocardiography in all patients. In 42 patients an additional imaging technique was performed during follow-up: CT scan in 19 patients, cMRI in 16 patients and 21 catheterizations. A second interventional procedure was indicated in 19/42 (45.2%) patients: 9 stent re-dilation, 7 new stent implantation and both procedures in 3 patients. Complications were more frequent in patients in which the stent was implanted at a younger age (< 14 years) (73% vs 27% p< 0.001). Stent fractures were diagnosed in 6/42 patients (14.3%), at a median time of 4.5 years (IQR 2.50-6.20) after stent implantation (range 2.1-12.5 years). Stent fractures were only visible in CT scan or fluoroscopy, not in CMR. Aneurisms in the coarctation site were diagnosed in 5 patients (11.9%).
Conclusions: Although stent implantation as treatment for aortic coarctation is an effective, feasible and safe technique, 45.2% of the patients will require an intervention during mid-term follow-up. Fluoroscopy and CT scan with 3D reconstruction should be incorporated in the follow-up of aortic coarctation patients after stent implantation.

MP1-4
Long term follow up of covered stents in the management of coarctation of the aorta

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Objectives: To evaluate the results of covered Cheatham-Platinum stent implantation in the management of native coarctation of the aorta and report long-term follow-up.
Background: Covered stents are being used increasingly in severe and complex coarctation of the aorta mainly to reduce risk of aortic wall complications. There is, however, limited data on the long-term outcome.
Patients and Methods: Eighty-six patients receive 90 covered Cheatham-Platinum stents (January 2002–December 2013) at a single center—in 84 patients as primary treatment and in 2 as a rescue. Mean age was 21.56 (11–56) years and mean weight 56.5 (30–102) kg. Primary end points were reduction in systolic pressure gradient and an increase in coarctation segment diameter. Changes in antihypertensive medicines and complications were recorded on follow-up.
Results: Mean coarctation segment diameter increased from 4.40 to 16.32 mm (P< 0.0001). The systolic gradient decreased from mean of 54.63 to 4.4 mm Hg (P < 0.0001). There was one death related to anaesthesia, 3 days post procedure due to cerebral anoxia. There was one dissection diagnosed 24-hr post procedure. At a mean follow-up of 85.9 (56–144) months, all stents were patent and in good position on computed tomography. Six (7%) patients underwent successful re-dilation. Antihypertensive medication was decreased or stopped in 51 (59%) patients.
Conclusions: Covered Cheetham-Platinum stents are an effective form of therapy in selected patients with complex and severe coarctation of the aorta. Aortic wall complications can occur even with covered stents. Covered stents are a safe alternative to conventional stenting in the long-term. They can be re-dilated safely to keep pace with somatic growth.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

MP1-5

Percutaneous lymphangiosclerosis as treatment for protein loosing enteropathy and plastic bronchitis in patients with failing Fontan circulation

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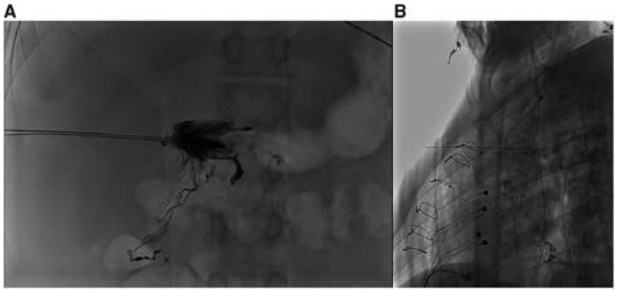
Objectives: To determine the feasibility and clinical short term results of superselective lymphangiosclerosis in Fontan patients with protein loosing enteropathy PLE and plastic bronchitis PB.
Methods: Dilated lymph vessels in periportal (PLE)(fig A) or paratracheal (PB)(fig B) position were punctured with a 22G Chiba needle; good intralymphatic position ascertained by water soluble contrast injection with drainage to abnormals lacteals; after flushing with glucose 5% (to evacuate most ions), occlusion of distal lymph vessels was obtained by injection of 1-3 cc of a mixture of lipiodol/ n-BCA N-butyl cyanoacrylate (Histoacryl®) 4/1 (which will solidify when in contact with ions as it spreads in lymph vessels). Effect on symptoms, plasma albumin or expectorations was monitored.
Patients & Results: 4 patients with persistent PLE were treated with periportal lymphangiosclerosis; Fontan at 3.4±0.4 years; PLE started 2.3±1.0 (range 1.0-3.3) y after Fontan; time of procedure since start PLE 8.2±3.7 (range 3.3-12.2) y; in all patients (1 patient required a 2nd procedure) the lymphangiosclerosis resulted in complete lasting normalisation of albumin levels after withdrawal of all medication (diuretics, steroids, pulmonary vasodilators) (FU 4-8 months). Symptoms of diarrea and abdominal bloating disappeared with significant improvement of quality of life.
1 pt (Fontan at 2.9 y; age 16.4y) with PB for 2 years had exacerbation of casts expectorations after a surgical procedure. Inguinal intranodal lymphangiography failed to improve symptoms, but demonstrated peritracheal dilated lymphatics. Direct puncture (left and right parasternal) with paratracheal lymphosclerosis resulted in lasting absence of tracheal casts (FU 3 months).
Conclusions: Periportal/peritracheal lymphangiosclerosis is a very promising technique in Fontan patients with PLE/PB. Larger series are needed to determine the incidence and reasons of success/failure, with long term results on lymph leak and effects on liver function.



MP1-6

Early postoperative cardiac catheterization in children: a single center retrospective survey over 5 years

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Background: Cardiac catheterization is a main tool in complicated postoperative course as a diagnostic or as a therapeutic modality. We report a retrospective single center series.
Material and methods: Between August 2013 and August 2018 all patients with postoperative cardiac catheterization within 30 days after surgery were included. Demographic, type of surgery, Artistotle score, reason for cardiac catheterization (group 1. treatment of postoperative lesions; group 2: treatment of associated abnormalities; group 3/ diagnostic procedure) were collected. Type of intervention, procedural complication, in hospital mortality were recorded.
Results: 164 patients were included. Median time between surgery and catheterization was 1 day. 36,5% were neonates. 24,3% were on ECMO. Mean age and weight were 16 months and 7,6 kg. Average of Aristotle surgical score was 9,5 and complexity level of cardiac catheterization was 3. Reason for catheterization were group 1: N=36, group 2 N=56 and group 3: N=72. 73 diagnostic procedures and 104 endovascular interventions in 91 patients were performed (17 atrioseptostomies, 13 balloon dilatations, 47 embolisations, 25 stent implantations; 1 vsd closure and 1 valvuloplasty). The precatheterization diagnostic was modified in 16,5% of patients (N=27/164). There were 4 post procedural surgeries: 3 because of modified diagnosis and 1 for procedural complications. 7,9% of complications and no per procedural death were observed. Mortality before discharge was 13,41%.
Conclusions: Early cardiac catheterization is mandatory for patients with complicated course after complex cardiac surgery. Change of diagnosis is not rare, endovascular treatment can be performed with low complications even in high risk patients.

MP1-7

Atrial septal defect closure (ASD and PFO) without fluoroscopy in both paediatric and adult patients – 19 years of experience

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Introduction: Defects of the atrial septum are very common congenital heart disease; closure might be indicated and performed interventionally with transcatheter devices implantation guided by fluoroscopy. One big disadvantage of transcatheter procedure is the x-ray exposition and contrast agent exposure of the patient and examiner. So the aim of this study was to clarify whether interventional closure of atrial septal defects is possible and safe when guided by transesophageal echocardiography (TEE) alone.
Methods: This is a retrospective single centre study of all paediatric and adult patients undergoing interventional atrial septal defect (ASD) or persistent foramen ovale (PFO) closure without fluoroscopy during 1999-2018.
Results: 758 / 2357 (37%) patients were included with interventional ASD or PFO closure with all complexity by transesophageal echocardiography (TEE) only. The rate of patients without fluoroscopy was low (8-32%) in the early decade (1999-2009) but has now increased to 40-90% of the patients in the last decade (2010-2018). Closure were was performed with Amplatzer® Septal Occluder (ASO) for ASD II or PFO Occluder (APO), Gore Cardioform PFO occluder, Ceraflex-ASD and Figulla Flex ASD. Interventional procedure succeeded in 92% and initial (day of intervention) closure rate was 93.6%. 50% were patients < 18years. Complication rate and examination time was quite similar to the usual procedure with fluoroscopy. In only 49 / 2357 patients (2.1%) procedure was switched from TEE alone to fluoroscopy due to difficulties of controlling wire or device in the beginning decade.
Conclusion: Interventional closure of ASD is safe and effective in paediatric and adult patients without fluoroscopy and with

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

transoesophageal echocardiography (TEE) alone. Therefore TEE guided closure of ASD and PFO should be considered in order to avoid unnecessary radiation exposure for all but especially for the paediatric patients.

MP1-8

Time and method of diagnosis of severe congenital heart defects

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Background: Congenital heart defects (CHD) are the most common birth defects worldwide and are an important cause of morbidity and early death. The time of diagnosis may have impact on treatment and outcome. The objective of this study was to investigate the time and method of diagnosis in children with severe CHDs.
Methods: Data concerning all pregnancies and children with severe CHD in Norway in 2016 were retrieved from the Norwegian Registry of Pregnancy Termination and the Oslo University Hospital's Clinical Registry for Congenital Heart Defects.
Results: In this nationwide register-based cohort study, which included all 60528 live births, stillbirths and late terminated pregnancies in Norway in 2016, 181 (0.3%) fetuses with severe CHDs was identified. The severe CHD was identified before birth in 105 (58%) fetuses. A total of 51 (49%) pregnancies with severe CHD were terminated. In most children without prenatal diagnosis, the CHD was identified during the birth hospitalization, however, in nine (12%) children without a prenatal diagnosis, the heart defects were discovered after discharge from hospital. The method of CHD detection in live born children without prenatal diagnosis is presented in figure 1.
Conclusions: Most children with severe CHDs were detected by ultrasound examination during pregnancy or routine examinations prior to maternity leave. However, almost half of the children diagnosed with severe CHD after birth were found outside of routine examinations and in some children the diagnosis was not recognized until after hospitalization. The importance of immediate assessment by a cardiologist of small children with suspected severe CHD has to be emphasized

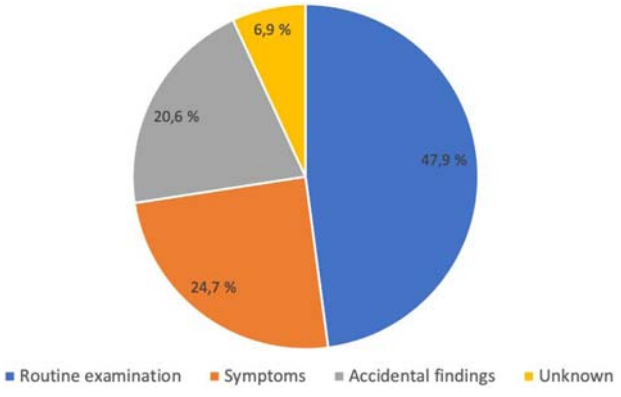

Figure 1.
The method of CHD detection in live born children without prenatal diagnosis

MP1-9

Wearable-based physical activity assessment in children with congenital heart disease

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Objectives: Still, physical activity (PA) is low in children with congenital heart disease (CHD). Wearables, validated devices to assess daily PA in children is used in this study to compare PA in CHD patients to healthy peers (step-count and moderate to vigorous physical activity (MVPA)) and whether they follow the WHO criteria of at least 60 minutes of per day.
Methods: From September 2017 to September 2018, 84 young patients (11.0 ± 3.9 years, range: 5.9-17.6 years, 32 girls) with various congenital heart diseases (CHD) participated in a wearable-based PA assessment, recorded with the Garmin vivofit® jr for seven consecutive days. Step-count and MVPA were calculated and compared to a healthy reference cohort (RC) of 86 children (11.0 ± 3.9 years, 45 girls) via Student's T-test for independent samples.
Results: Children with CHD showed a lower step-count (CHD: 10,052 ± 3,354 steps/day vs. RC: 11,822 ± 3,635 steps/day, p<.001) and lower MVPA (CHD: 73.4 ± 24.9 MVPA minutes/day vs. RC: 84.2 ± 25.1 MVPA minutes /day, p=.010) compared to healthy peers. Comparisons of MVPA throughout the week highlighted no significant difference during weekdays while on the weekend CHD reached lower levels of MVPA (CHD: 64.4 ± 30.0 MVPA minutes /weekend-day vs. RC: 78.6 ± 32.6 MVPA minutes/weekend-day, p=.003). According to WHO criteria, 71.4% of CHD reached the recommended 60 minutes MVPA per day on a weekly average.
Conclusions: The current study shows that children with CHD still have a lower step-count and MVPA in daily life compared to their healthy peers. Nevertheless, though the majority is sufficiently active, an active lifestyle still needs to be promoted.

MP1-10

Maternal diabetes as a risk factor for high blood pressure in late childhood: a prospective birth cohort study

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Intrauterine fetal conditions can have lifelong cardiovascular effects. The impact of maternal diabetes on children cardiovascular profile is not well-established. The goal of this study was to examine the association between maternal diabetes and offspring's blood pressure up to 10 years of age and to explore BP trajectories in this age range. Generation XXI is a prospective birth cohort which enrolled 8301 mother-offspring pairs, including 586 (7.1%) children of diabetic mothers. The associations between maternal diabetes and blood pressure at 4, 7 and 10 years of age was modeled using linear regression. A mixed-effects model was built to assess differences in blood pressure variation over time. Path analysis was used to quantify effects of potential mediators.
Maternal diabetes was associated with higher blood pressure in offspring at age of 10 (systolic blood pressure: β 1.48, 95% CI: 0.36, 2.59; diastolic blood pressure: β 0.86, 95% CI: 0.05, 1.71). This

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

association was independent of maternal perinatal characteristics and it was partly mediated by child's body mass index and, to a lesser extent, by gestational age, type of birth and birthweight (indirect effect proportion 73%). No significant differences in blood pressure levels were found at 4 and 7 years-old. Longitudinal analysis showed an accelerated systolic blood pressure growth on maternal diabetes group (β 1.16, 95% CI: 0.03, 2.28). These finding were especially relevant in males, suggesting sex differences in the mechanisms of blood pressure prenatal programming. Our results provide further evidence that maternal diabetes is associated with higher blood pressure late in childhood, demonstrating a significant role of child's body mass in the pathway of this association.

MP1-11

Updated and revised reference values of aortic pulse wave velocity in children and adolescents aged 3-18 years

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Introduction: Measurement of aortic stiffness – expressed as aortic pulse wave velocity (PWVao) – is an accepted method in the process of detecting organ damages and stratifying individual cardiovascular (CV) risk in adults. Diseases in children and adolescents might influence aortic stiffness. It is necessary to exclude overweight (OW), obese (O) subjects, and individuals with increased systolic (SBP) and/or diastolic blood pressure (DBP) from the population, when creating reference values of PWVao in the pediatric population. Body mass index (BMI), SBP and DBP cut-off values have changed in this population during the last decade.
Aims of our study were to expand the database of our previously published (2012) reference values of PWVao for children and adolescents; and to revise it by the application of the recently determined BMI and SBP, DBP cut-off values.
Methods: PWVao was measured by an invasively validated occlusive-oscillometric device in a healthy population aged 3-18 years. To categorize subjects into OW and O subgroups, cut-off values published by Cole (2012) were used. Increased SBP, DBP were defined by applying the reference values published by Schwandt (2015). Finally, n=4.690 (2.599 boys) participants were recruited.
Results: Mean PWVao values increased with around 1 m/s between the ages of 3 and 18 years in both sexes, namely, this parameter rose from 5.4±0.6 to 6.4±0.5 m/s ($p<0.05$) in boys and from 5.5±0.6 to 6.4±0.5 m/s ($p<0.05$) in girls. Mean PWVao values were significantly higher in boys according to girls in the age groups of 13-15, and 17 years. After the comparison of mean PWVao values measured in 2012 and 2018, significantly lower mean PWVao values were found in 2018 (in boys in the age group of 7-16 years; in girls in the age group of 10-17 years).
Conclusions: To the best of our knowledge this is the largest database of PWVao of the healthy population aged between 3-18 years published to date. Due to the change of anthropometric and physiological cut-off values during the last decade, the "old" database of PWVao needed to be revised. As a result of this, reference values of PWVao decreased significantly in both genders.

MP1-12

Reduced handgrip strength is associated with lower health related physical fitness in children with congenital heart disease

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Introduction: Technical development in surgery and an enhancement in cardiology care have led to a decrease of the mortality rate in children with congenital heart disease (CHD). At the same time, functional outcomes like health-related physical fitness (HRPF) did not improve as well. Handgrip strength as a simple and effective method to identify adults at higher risk of cardiovascular mortality could be a useful tool to identify young CHD patients at risk for reduced HRPF.
This study aimed to assess handgrip strength and HRPF in CHD and compare their results to a healthy control group (CG).
Methods: Starting in May 2014 until October 2018 handgrip strength and HRPF were assessed in 304 patients (12.5 ± 3.4 years; 117 girls) with various CHD (69 simple; 89 moderate and 146 complex severity). Results were compared to 1560 children from a CG (11.5 ± 2.7 years; 925 girls) recruited in a school project in 2016. Handgrip strength was calculated for the dominant hand (maximum value from 3 repetitions of the right and 3 of the left hand). HRPF was tested by five FITNESSGRAM® motor tasks of the and converted into standard deviation scores (SDS) according to the values of CG.
Results: After adjusting for sex and age patients with CHD showed significantly lower handgrip strength in comparison to the CG (CHD: 19.6 ± 5.6 kg; CG: 24.4 ± 5.6 kg; $p<.001$) and significant lower HRPF (SDS: -0.56± 0.82; $p<.001$). Children with complex CHD had lower values in handgrip strength compared to children with moderate (complex: 17.7 ± 5.7 kg; moderate: 19.0 ± 5.6 kg; $p<.001$) and simple (complex: 17.7 ± 5.7 kg; 23.7 ± 5.6 kg; $p<.001$) severity. Patients with total cavopulmonary connection had the lowest values (16.6 ± 5.6 kg). There was a significant relationship between handgrip strength and HRPF ($r= .316$; $p<.001$).
Conclusions: Children with CHD have considerable lower handgrip strength and HRPF compared to healthy children. The positive relation between both parameters highlight the potential of the handgrip measurement as a useful screening tool in the clinical context.

MP2-1

Additional antegrade pulmonary blood flow after partial cavopulmonary connection – impact on hemodynamics and pulmonary artery branch growth. A 14-year single center experience

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Background: Partial cavopulmonary connection (PCPC) is a widely used procedure for providing pulmonary blood flow in patients with a functional single ventricle. Controversy exists on whether additional antegrade pulmonary blood flow (AAPBF) is beneficial after PCPC operation. This study aims to investigate the impact of AAPBF after PCPC on hemodynamic parameters and pulmonary artery branch growth.
Methods: Medical files of patients who underwent PCPC between 2004 and 2017 were reviewed. Patients with missing data were excluded from the study. We divided the patients with PCPC in two groups according to AAPBF: group 1- patients with AAPBF, group 2 - patients without APBF. Cardiac catheterization data immediately before total cavopulmonary connection (TCPC) were analyzed and compared between the two groups. Data were presented as medians with range or as means ± standard deviation.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

A non-parametric Mann-Whitney U test integrated in the statistical software SPSS 24.0 was used. A value of P< 0.05 was considered significant.
Results: 77 patients were operated for 14 years. 71 patients were included in the study. Median age at PCPC was 10 months (2-44).In 34 patients (48%) AAPBF was preserved (group 1) and in 37 patients (52%) AAPBF was interrupted (group 2). There was no statistical significance between group 1 and group 2 concerning hemodynamic measurement just before TCPC - oxygen saturation 81% (63-93) versus 79% (65-93), pulmonary artery pressure 14 mmHg (7-23) versus 13 mmHg (8-17), pulmonary vascular resistance 1.12 Wood units (0.47-2.24) versus 1.34 Wood units (0.41 – 3.9), ventricular end-diastolic pressure 10 mmHg (4-18) versus 10 mmHg (6-16), ejection fraction 62% (47-87) versus 59% (45-85). Concerning the pulmonary artery growth, there was a statistical significance between group 1 and group 2 in left pulmonary artery dimensions with z-scores 1.57 versus 0.27 (p=0.001), with no statistically significant difference in right pulmonary artery dimensions with a z-score 0.83 versus 0.67.
Conclusion: Our data suggest that preserving the AAPBF at PCPC stage, has no adverse effect on hemodynamics at the time of Fontan completion. Moreover, there was an increase in pulmonary artery growth in the presence of AAPBF.

MP2-2
Faecal calprotectin levels as a diagnostic marker for necrotising enterocolitis in infants with congenital heart disease: cross-sectional validation study (preliminary data)
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Introduction: Infants with duct-dependent circulation defects experience suboptimal splanchnic perfusion resulting in gut inflammation which is exacerbated by cardiac surgery and enteral feeding, increasing the risk of necrotising enterocolitis (NEC). NEC leads to extensive feed interruptions contributing to poor growth particularly during the early post-operative phase but continuing up until discharge. Poor growth and longer hospital stay are risk factors for death in infants with congenital heart disease. Calprotectin (36.5kDa) is a neutrophil activation marker which is exhibited in the cytoplasm of neutrophils and expressed on activated monocytes and macrophages; and participates in leukocyte interactions with the endothelium and cellular adhesions, leading to the recruitment of leukocytes to inflamed intestinal tissue and hence a measure of gut inflammation (normal faecal calprotectin level <50mg/kg).
Methods: To date, faecal calprotectin levels have been measured in twenty term infants with duct-dependent circulation defects using ELISA methods. Samples are collected post-surgery once feeding is established and bowels open. Infants follow a high risk feeding protocol (starting at 0.5ml/kg) consisting of either expressed breast milk or hydrolysed formula (Pepti-junior). Infants with suspected NEC underwent an abdominal radiography.
Results: Incidence rate of NEC (Bell's stage 2) was 15%. The mean faecal calprotectin level for infants with NEC was significantly higher 3957mg/kg compared to those without 713mg/kg (95% CI 223, 6918; p-value 0.02). Infants with single ventricles have higher baseline calprotectin levels 2184mg/kg compared to other duct-dependent circulation infants (p-value 0.04). Time to establish 100ml/ kg feed volume was on average 10 days longer in the NEC group (22 days) compared to those without NEC.
Conclusion: Infants with duct-dependent circulation defects have elevated baseline faecal calprotectin levels, particularly infants with single ventricle. Faecal calprotectin may be a useful marker for gut inflammation eliminating abdominal radiography and exposure to radiation.

MP2-3
Extracorporeal membrane oxygenation (ECMO) in patients with hypoplastic left heart syndrome: a retrospective cohort study
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Introduction: Extracorporeal membrane oxygenation (ECMO) has been used with increasing frequency to support pediatric patients after repair or palliation of congenital heart disease. 10-12% of the newborns undergoing Norwood procedure require advanced circulatory support. The purpose of this study was to report our experience with functional single ventricle patients who were supported by ECMO after Norwood surgery.
Methods: In this retrospective cohort study, we enrolled patients with hypoplastic left heart syndrome (HLHS) who required ECMO support after a Norwood operation between July 2015 and June 2018. We evaluated demographic variables (age, weight, sex, presence of aortic atresia, presence of mitral atresia, ascending aorta diameter, and atrial septal defect diameter) and ECMO-related variables (local of ECMO initiation, indication and time under support). Then, findings were compared between survivors and nonsurvivors.
Results: A total of 21 patients were included in the present study. The median age of patients was 4 days (range, 2-69), the median weight was 3000g (range, 2600-3800), and 52.4% of patients were male. Aortic atresia was present in 52.4% of patients while mitral atresia in 47.6%. For 19% of patients, ECMO was initiated in the operation room (OR); for all other patients, in the intensive care unit (ICU). Indications for ECMO installation included cardiac arrest (57.1%), low cardiac output state (LCOS) (38.1%) and arrhythmia (4.8%). The median time under ECMO support was 8 days (range, 3-44) and the median follow-up time was 35 days (range, 4-917). Overall survival during the follow-up time of the study was 31.3% and none of the independent variables related to patients' demographics differed between survivors and nonsurvivors. All patients submitted to ECMO support above 9 days died and the survival rate for patients submitted to ECMO due to cardiac arrest (CA) was 12.5%, while it reached 53.3% for those undergoing ECMO for other reasons than CA (Log-rank test, p=0.05).
Conclusion: Although ECMO has been increasingly used to support patients with HLHS after stage 1 palliation, the mortality associated with the procedure is still high. Early identification and management of LCOS, before it progresses to cardiac arrest might reduce the mortality until hospital discharge.

MP2-4
Feasibility, usefulness, function and durability of a pulmonary valved conduit evaluated in a growing, long-term lamb model
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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Objectives: We evaluate a tissue engineered valved conduit connecting the right ventricular outflow tract to the pulmonary artery bifurcation during long-term follow up and growth up to 24 months.
Methods: In total, 19 female Swiss white mountain lambs (27–38 kg at surgery) were operated, using left lateral thoracotomy and cardiopulmonary bypass. The native pulmonary valve and the complete pulmonary trunk were resected. The conduit was constructed out of a de-cellularized porcine small intestinal submucosa extracellular matrix biologic scaffold and was implanted in orthotopic position. Follow-up transesophageal/-thoracic 2D Doppler echocardiography and laboratory values (LDH, hemoglobin, hematocrit, white blood cell count) were performed directly after surgery and after 1,3,6,12,15,18 and 24 months.
Results: Seven animals died perioperatively, 3 animals died due to asphyxiation pneumonia (at postoperative day 23) and infective endocarditis (day 20 resp. 256). In 9 animals, long-term function of the implanted conduit was assessed. Mean follow-up time so far is 17 months (range 20 days – 24 months). Seven animals were already sacrificed at 24 months (n=4), at 15 months (n=1), 12 months (n=1) and 9 months (n=1). Mean body weight at last follow up was 58.8 kg (28-72). 2D echocardiography revealed good function of the conduit with mean±SD systolic pressure gradient 7.1±1.3 mmHg and maximum moderate regurgitation. LDH increased from a pre-operative 976±78.2 to 1100±181.8 U/l [n.s.]. CT scan prior to termination (n=7) revealed no severe calcification or dilatation of the conduit. Subsequent histologic evaluation revealed a variable degree of incorporation to native tissue between individual valve leaflets. Most valves were moderately populated by stromal cells and showed endothelialisation (confirmed by CD31 immunohistology) and mild inflammation within the valve. Small foci of chondroid and osseous metaplasia were occasionally observed, predominantly at the suture sites rather than within the leaflets.
Conclusions: The implantation of a valved RV-PA-conduit in a growing lamb model is feasible, useful and leads to a stable valve function up to 2 years. The animals demonstrated, after the initial recovery phase from surgery, good physical development but remain at risk for endocarditis (16%). The function of the valved conduit was satisfactory during long-term follow up.

MP2-5
The Influence of cardiac surgery on intestinal perfusion in children with congenital heart disease

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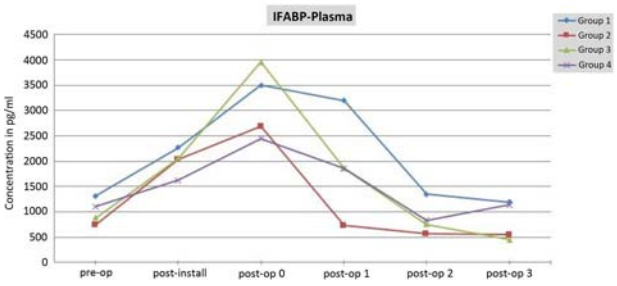
Introduction: Intestinal perfusion is impaired in children with congenital heart disease (CHD) depending on the hemodynamic status. Especially newborn with duct dependent pulmonary or systemic perfusion are at risk for severe intestinal complications before and after cardiac surgery due to bad perfusion or hypoxic state. Intestinal fatty acid binding protein (IFABP) can be detected in plasma and urine only after cell damage of the intestine. Therefor it is a biomarker for intestinal injury that can be detected in Plasma or urine. To understand the influence of heart failure to intestinal perfusion we analyzed perioperative samples from children with different heart defects.



Methods: We investigated plasma and urine samples from 81 children (mean age 108 d) with 104 surgical events pre- and postcardiac surgery to understand the influence of hemodynamic changes to intestinal perfusion analyzing IFABP. Samples were collected before surgery (pre-OP and post-install), right after surgery (post-OP) and at day one to three after surgery (post-OP 1-3). Depending on the heart defect and hemodynamic status we subdivided the patients in 4 main groups (1=Duct/Shunt dependent Defects with Norwood-Type surgery and/or BT-Shunt, TGA; 2= Stage 2 Palliation {Glenn}, TOF-Repair; 3= Coarctation of the aorta {ISTA}, 4= stable hemodynamic status atrial septal defect, ventricular septal defect, pink Fallot).
Results: IFABP can be detected in plasma and urine samples with a good correlation. All Patients show a peak after surgery (plasma and urine). Group 1 and 3 (newborn) start with a higher burden, showing the highest level overall after surgery (3494 pg/ml and 3953 pg/ml). G3 level is higher in post-OP-serum but recovers quicker compared to the shunt dependent defects. Due to short halftime in blood, urine IFABP is higher post-OP.
Conclusions: Surgical repair of CHD always leads to cell damage of the intestine. IFABP is a good biomarker for indicating intestinal damage due to poor perfusion in children with congenital heart disease. Duct or shunt dependent hemodynamics seem to lead to a higher burden and might indicate a higher risk for intestinal complications. To evaluate the clinical relevance further investigation needs to be done.

MP2-6
Surgical Valvuloplasty in Neonates and Infants with Congenital Aortic Valve Stenosis

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Objectives: In terms of new surgical techniques, congenital aortic valve stenosis treatment is under controversy. This study sought to retrospectively analyze surgical valvuloplasty effectiveness in neonates and infants.
Methods: From July 2012 to February 2018, 84 consecutive neonates (27 patients ≤30 days) and infants (57 patients at age ≥31 days and <1 year) were included. 9 patients (10.7%) underwent a preceding balloon valvuloplasty. The indications for the procedure were stenotic disease. Endocardial fibroelastosis was present in 4 patients (4.8%). The procedures performed were subaortic stenosis repair (n = 5; 6%), commissurotomy (n = 74; 88%), leaflet shaving (n = 64; 76%), raphe shaving (n = 15; 18%), raphe resection (n = 17; 20%), and leaflet replacement (n = 3; 3.5%). Post-repair geometry was tricuspid in 28 (32%) patients.
Results: The survival rate was 100%. Freedom from re-repair and Ross operation at 5 years was, respectively, 96.4% (95% confidence

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

interval) and 94% (95% confidence interval). In multivariate analysis, previous balloon dilatation before 6 months of age, the absence of a developed commissure, cusp retraction, a non-tricuspid post-repair geometry and cross-clamp duration were predictors for redo operations. After a mean follow-up period of 2.9±1.6 years, 76 (90.5%) patients had a preserved native valve, with undisturbed valve function (peak gradient <40 mmHg, regurgitation <mild) in 45 (53.6%), whereas 20 had moderate regurgitation and 19 had moderate stenosis.
Conclusions: Surgical valvuloplasty is safe and durable allowing the patient to grow to infancy and, ideally, into adulthood when aortic root stabilization is available and re-repair or Ross operation can be performed with excellent results or to prevent replacement. Avoidance of early balloon dilatation and aiming for a tricuspid post-repair arrangement, appropriate definitive solution regarding cusp retraction may improve outcomes.

MP2-7

Mid-term clinical outcome after Fontan conversion compared with primary total cavopulmonary connection.

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Background: The indication of Fontan conversion from atriopulmonary connection (APC) to total cavopulmonary connection (TCPC) is unclear.
Methods: In order to analyze mid-term outcome after Fontan conversion, TCPC patients who underwent cardiac catheterization at > 18 years of age between July 2005 and November 2018 were included and divided into two groups according to the first Fontan surgery (APC group, n=27; TCPC group, n=22).
Results: Fontan conversion in APC group was undertaken at the mean age of 22.2 ± 5.3 years. Ten cases (38%) had atrial tachyarrhythmia before Fontan conversion. Antiarrhythmic surgery was added in 25 cases, where 6 cases (24%) developed sinus node dysfunction after the procedure. Four of them required pacemaker implantation. Cardiac catheterization at the mean age of 27.4 ± 6.3 years in APC group and 26.2 ± 6.1 years in TCPC group showed no significant difference in SVC pressure (12.6 ± 2.5 mmHg vs 13.5 ± 4.1 mmHg), ventricular end-diastolic pressure (10.9 ± 3.5 mmHg vs 11.6 ± 5.1 mmHg), and cardiac index (3.0 ± 0.9 L/min/m2 vs 2.5 ± 0.4 L/min/m2) between two groups. Median serum BNP at the time of catheterization was similar 16.4 pg/mL vs 21.6 pg/mL. Recurrence of tachyarrhythmia was seen in one in APC group immediately after surgery. Two patients in TCPC group newly developed atrial flutter.
Conclusions: Fontan conversion with antiarrhythmic surgery seemed to control future atrial tachyarrhythmia, though a few cases were complicated sinus node dysfunction. Hemodynamics and ADL were similar between two groups in their early 30s.

MP2-8

Valve prosthesis in the right ventricular outflow tract: An analysis of the German National Register for Congenital Heart Defects

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Background: Replacement of the pulmonary valve is required for native valve vitals or after pulmonary artery intervention. The durability of the prostheses (homo- and heterografts) is limited for several reasons in children, adolescents and adults. This necessitates replacement of the prostheses during the course of life and thus re-operations / interventions are needed. The choice of operation time and type of prosthesis is therefore of great importance to the patient and the attending physician.
Methods: The starting point for the present data analysis is the National Register for Congenital Heart Defects (NRCHD). The NRCHD currently has more than 53,000 patients with congenital heart disease (CHD) registered. The database was systematically screened for patients undergoing pulmonary valve intervention. In total, 1,202 patients were identified who had a native pulmonary valve vitium [Fallot's tetralogy (TOF), pulmonary valve stenosis (PS)] or re-pulmonary-valve-vitium [e.g. after ROSS-OP, after arterial switch surgery or after correction Truncus arteriosus (TAC)], pulmonary valve replacement and prosthesis types (homograft, heterograft or mechanical prosthesis), size and type of valve.
Results: In the 1,202 patients (526 (43.8%) female, 676 (56.2%) male) the mean age was 25 ± 12.8 (minimum 2 years, maximum 84 years). The three most common primary cardiac diagnoses are TOF (53.6%), TAC (14.7%) and congenital aortic valve stenosis (9%). The most commonly used prostheses for pulmonary valve replacement were the aortic homograft (33.6%), the Contegra valve (32.7%), and the interventional pulmonary valve replacement with a melody valve (18.6%). Only 1% of patients received a mechanical valve replacement. In total, 2,132 surgical and/or interventional pulmonary valve replacements were documented in 1,202 patients. 632 patients received more than one prosthesis. The TOF patients were those with the highest number of replaced pulmonary valves (644 interventions), followed by 177 interventions in patients with TAC and 108 interventions in patients with congenital aortic valve stenosis.
Conclusion: Overall, many interventions/operations were made with replacement of the pulmonary valve. Most patients get the first surgical valve replacement in childhood. The effects of the later required surgical and interventional re-interventions/operations on the individual disease process must be further investigated.

MP2-9

Serum levels of growth differentiation factor 15 are associated with outcomes in patients with a Fontan circulation

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Introduction: Growth differentiation factor 15 (GDF-15) is involved in noncardiac and cardiac stress pathways. Elevated levels are associated with mortality in acquired heart disease. The aim of this study was to investigate GDF-15 in Fontan patients.
Methods: In this prospective study, Fontan patients were followed at the University Medical Center Groningen from 2012 to 2018. Serial serum GDF-15 measurement and clinical assessment was done both at baseline and after two years. The association between GDF-15 and clinical outcome, including Fontan-related hospitalization and all-cause mortality, was investigated.
Results: Eighty-one patients were included, of which 51 patients had a 2-year follow-up. Median age at baseline was 21 years (IQR 14.5-27.5) (table 1). Median GDF-15 levels at baseline were 552 pg/mL (IQR 453-729). Patients in NYHA class III (n=10) had higher GDF-15 levels than patients in NYHA class I or II (n=71) (1086 pg/mL (IQR 659-1239) versus 539 pg/mL (IQR 443-670), P=0.001). GDF-15 correlated positively with age and time since Fontan completion, Fontan type, γGT and beta blocker use and negatively with exercise capacity. There was no significant relationship with sex, ventricular ejection fraction or morphology. During a median follow-up of 58 months (IQR 39–66), the clinical outcome occurred in 33 patients (41%): 30 Fontan-related hospitalizations and 3 deaths. Patients with an elevated baseline GDF-15 (n=20, defined as upper quartile) had a higher risk of hospitalization or death (HR 3.8, 95% CI 1.9-7.5, P<0.001). This relationship persisted after individual and multivariate adjustment for independently predictive covariates (HR 3.0, 95% CI 1.5-6.2, P=0.004). Median
GDF-15 did not increase significantly after 2 years in patients with serial GDF-15 measurements (570 pg/mL (IQR 450-740) versus 595 pg/mL (IQR 483-872), P=0.146). Patients with a GDF-15 increase >70 pg/mL (n=13, defined as the upper quartile of interval change between baseline and the second visit) had a higher, yet not statistically significant, risk of hospitalization or death (HR 2.3, 95% CI 0.9-5.8, P=0.071).
Conclusions: In Fontan patients, elevated serum levels of GDF-15 are associated with worse functional status and increased risk of Fontan-related hospitalization or death. The additional value of serial GDF-15 measurements requires further investigation.

Table 1- Baseline characteristics (n=81)

Age (years)	21.0 (14.5-27.5)
Females	41 (51)
Left ventricular morphology	68 (84)
Time since Fontan (years)	15.4 (9.0-22.5)
Type Fontan	
Atriopulmonary connection	16 (20)
Lateral tunnel	44 (54)
Extracardiac tunnel	21 (26)
NYHA class	
I/II	71 (88)
III	10 (12)
Ejection fraction (%)	58.0 (50.4-62.3)
ß-blocker users	20 (25)
γGT (U/L)	59.2 (39.4-102.5)
Peak VO_2 (ml/min/kg)	23.9 (19.5-32.7)

All values are expressed as median (interquartile range) or frequency (percentage)

Table 1.

Pt	Age-months	Diagnosis	Device	Days on VAD	Pre-VAD FE	EF at weaning	Post-Weaning EF	Outcome
LG	120	IDCM	BH 25cc	210	8%	53%	Failed	HTx
PM	23	IDCM	BH 10cc/ Levitronix	21	15%	55%	50% (468 days-FU)	Weaned
AI	29	IDCM	BH 25cc	296	16%	45%	40%	Weaned (died after LVAD Explantation)
MG	33	IDCM	Levitronix/BH 15cc/Infant Jarvik 2015	180	18%	50%	Failed after 7 days	On VAD
RA	8	Myocarditis	BH 15cc	210	15%	45%	40% (111 days-FU)	Weaned

MP2-10
Weaning in Children with Left Ventricular Assist Device Support: A Single Center Experience

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Objective: To review our experience in pediatric patients that underwent LVAD placement and were selected for weaning after evidence of relevant and stable cardiac improvement on LVAD as measured by seriate echocardiographic evaluation.
Methods: From 2007 to 2018, a total of 55 LVAD were implanted at our institution. Longitudinal echocardiographic data were available for the analysis in 31 children who received LVAD implantation during the study period with a diagnosis of dilated cardiomyopathy and severe HF. Standard 2D and 2D Speckle Tracking-derived Echo measurements were evaluated at baseline, after LVAD placement and weekly until LVAD explantation and/or last follow-up. Patients were considered eligible for weaning if LVEF had improved during LVAD support and ≥45% with a remodeling of LV in terms of Z-score diameters. Patients eligible for weaning underwent a pump off trial.
Results: Five (9%) children were considered eligible for LVAD weaning. Median age was 29 months (IQR 23-33), median weight was 13 kg (IQR 11-15.2), with a median duration of support of 214 days (IQR 210-296). Myocardial biopsies and virus assessment excluded myocarditis in 4 cases, with 1 positive endomyocardial biopsy (EMB) for myocarditis and positive parvovirus B19 serology and BEM. Three pts were successfully weaned by LVAD, with 2 failure (one during the pump off trial and the other after 7 days) (Table 1).
Conclusions: LV recovery in children on VAD is possible although the reported rate of cardiac recovery is low. Serial Echocardiographic parameters are needed for the detection of myocardial improvement prior to attempting device explantation. Future and multi-center investigations are needed to define the clinical predictors of cardiac recovery which are still lacking in pediatric heart failure.

MP2-11
Airway Narrowness Affects the Severity and Onset of Respiratory Symptoms in Patients with Vascular Ring

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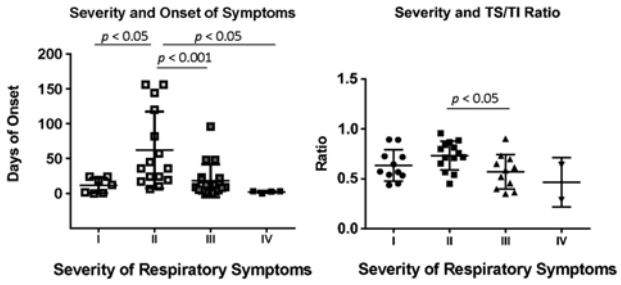
Introduction: Vascular ring is an uncommon congenital aortic arch anomaly where trachea and esophagus are compressed by the surrounding vascular structures and cause progressive respiratory

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

and/or esophageal symptoms during infancy and childhood. Variable degree of respiratory symptoms has been attributed to the tightness of the ring, but the relationship between the two has not been established. We tested whether the degree of tracheal obstruction correlates with the severity of respiratory symptoms in patients with vascular ring.
Methods: We retrospectively reviewed 61 patients with isolated vascular ring who underwent surgical repair and studied their clinical presentation and the anatomical size of trachea at the time of diagnosis. The respiratory symptoms were classified as I) no symptoms (n = 19), II) mild symptoms (cough, snoring, loud breathing, or stridor; n = 21), III) moderate symptoms with respiratory functional abnormalities (increased work of breathing or recurrent respiratory infection; n = 17), and IV) life threatening conditions (cyanosis, apnea, or cardiopulmonary arrest; n =4). The airway diameter was measured by either CT or MRI. Tracheal narrowing was assessed by the ratio of the narrowest tracheal diameter (TS) to the narrowest diameter at the thoracic inlet (TI).
Results: The onset of symptoms was earlier (median of 11.5 weeks) in IV compared to 9 months and 3 years in group III and II, respectively (both $p < 0.05$). Among symptomatic groups, there was weak positive correlation between the severity of symptoms and the degree of tracheal stenosis. Post-operative hospital days tended to be longer in patients with severe symptoms (II: 3.3 ± 1.2 days, III: 6.3 ± 5.8 days, and IV: 8.0 ± 2.0 days). Group I showed the widest variation of the all groups.
Conclusions: Although the degree of tracheal narrowing did not correlate with either onset or severity of symptoms, patients with severe respiratory symptoms tended to become symptomatic earlier. There may be other factors contributing to the severity of respiratory symptoms.



MP2-12
Myofiber organization in the failing systemic right ventricle of congenital heart diseases
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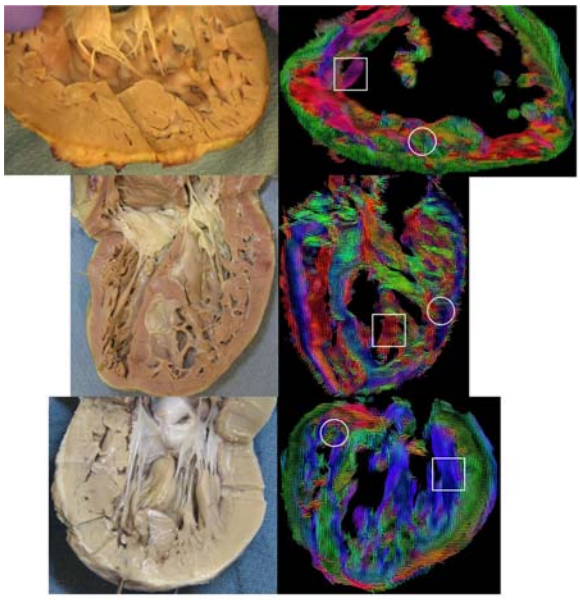
Basis: Heart failure and arrhythmia are common complications in patients with a systemic right ventricle. Magnetic resonance imaging (MRI) studies have shown microstructural alterations in left ventricular myocardium of hearts affected by genetic and acquired cardiac diseases. In this work, we tested the hypothesis that myofiber architecture is abnormal in the systemic RV of explanted, failing hearts affected by congenital diseases.
Methods: We used diffusion-weighted magnetic resonance imaging (DW-MRI) (3T Siemens Skyra) to examine 3 failing hearts explanted at transplantation from young patients (5-19 years) with a systemic right ventricle and a pulmonary left ventricle (1 case). DW MRI was acquired with a pulsed gradient spin echo sequence, single shot EPI readout and 1.5x1.5x1.5mm spatial resolution. Our gradient scheme contained 4 b=0s/mm2 images and 3 shells (b=1000, 1500, 2000s/mm2) of 30 gradients each. Nine averages were acquired with both AP and PA phase encoding directions. Diffusion compartment imaging was computed to separate the free diffusive component representing free water from an anisotropic component characterizing the orientation and diffusion characteristics of myofibers. The orientation of each anisotropic compartment was displayed in glyph format and used for qualitative description of myofibers and for construction of tractograms. Blocks of ventricular myocardium were removed for comparison with diffusion imaging.
Results: The hypertrophied systemic RV has an endocardial layer, comprising about 2/3 of the wall thickness, composed of hypertrophied trabeculae and an epicardial layer of circumferential myofibers somewhat like normal (Fig). Smaller trabeculae are organized with parallel fibers while larger, composite bundles show myofiber disarray, largely between component trabeculae. The apical whorl is disrupted and we observed myocardial whorls or vortices and abrupt fiber tract interruptions in multiple regions away from the apex. Histological exam of tissue blocks removed after imaging confirmed the diffusion imaging findings.
Conclusion: Myofiber organization is abnormal in the failing systemic right ventricle and might be an important substrate for heart failure and arrhythmia. It is unclear if the abnormal myofiber organization is due to hemodynamic factors or intrinsic developmental problems.



HLHS (top), CTGA (middle) and DIRV (bottom). Small trabeculae with coherent fibers (boxes) and vortexes (circles). Note marked disarray between trabeculae.

MP3-1
Evolution of NT-proBNP in the first 28 days of life in very low birth weight infants
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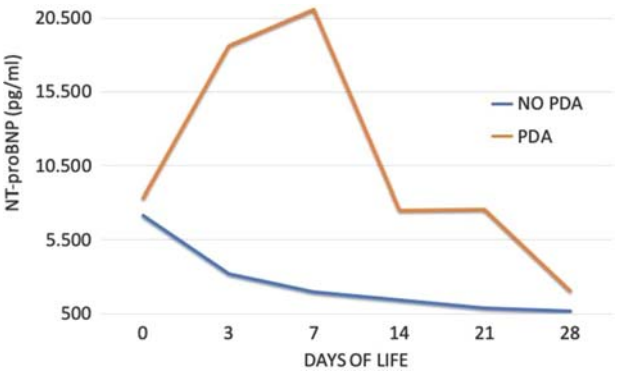
Objectives: To examine the evolution of plasmatic levels of N-terminal cerebral natriuretic propeptide (NT-proBNP) in the first 28 days of life of very low birth weight infants (VLBWI) both in the presence and absence of ductus arteriosus

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



Methods: Prospective study including VLBWI with birth weight ≤ 1500 grams and / or ≤ 32 weeks of gestational age (GA) admitted to the Neonatal Intensive Care Unit from 2015 to 2017. Weekly echocardiograms and biochemical determination of plasma NT-proBNP (pg/ml) were made during the first 28 days of life. NT-proBNP levels were longitudinally analyzed and correlated to echocardiographic parameters accounting for the presence of patent ductus arteriosus (PDA).
Results: We included 101 preterm with a mean GA of 28.85 weeks (± 1.85 SD) and mean birth weight of 1152 grams (±247.4 SD). A total of 139 NT-proBNP determinations were performed.
In the first 24 hours there is no difference in the plasmatic level of NT-proBNP in those patients who present with PDA 5,246 ng/dL (1,574 - 22,390) compared to those with no PDA 6,437 ng/dL (1,300 -14,444) (p = 0.49).
At 3, 7, 14 and 21 days of life patients with PDA had higher NT- proBNP values compared to those with no PDA. NT-proBNP plasmatic levels follow a different evolution in both groups: those VLBWI with PDA were found to have a progressive increase in NT-proBNP levels with a maximum peak at 7 days of life while in those with no PDA NT- proBNP levels decrease progressively with a nadir at 28 days of age (685.52 ng/dL). At 28 days of age we found no difference between both groups.
Conclusions: NT-proBNP levels in the first day of life and at 28 days of life cannot be considered a marker of PDA in VLBWI. At 7 days of life NT-proBNP levels
NT-proBNP levels differed the most in both groups. Importantly, in VLBWI with PDA, NT-proBNP levels also decrease over time.

MP3-2
Parameters of myocardial deformation in single ventricle patients after Fontan palliation – assessment by feature tracking cardiovascular magnetic resonance
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Introduction: Reliable quantification of cardiac function in single ventricle (SV) patients after Fontan palliation is challenged by complex cardiac anatomy and restricted acoustic windows. Feature-tracking (FT) cardiac magnetic resonance (CMR) is increasingly used in congenital heart disease. We sought to test the correlation between FT and conventional CMR-parameters, clinical parameters and biomarkers.
Methods: Myocardial deformation was retrospectively analyzed on cine-SSFP images in 40 consecutive patients (25 males) with SV physiology, 11±6 years after Fontan completion. Endocardial mid circumferential strain (mid-CS) and global longitudinal strain (GLS) were measured by FT (Qstrain, Medis Version 3.3) in short-axis and horizontal-long-axis, respectively. Image temporal resolution was <25msec.
Results: A dominant right ventricle (RV) was present in 16 (40%) patients, Fontan completion was performed at 3.2±1.5 years with a total cavopulmonary connection in 37 (93%) (18 fenestrated). Hospitalization length was 23±10 days, 16 patients experienced complications. Age at CMR was 15±7 years, weight 51±22 kg. Mean EDV was 89±33 ml/m2, ESV 41±18 ml/m2, EF 53±8%. At time of CMR, echocardiography showed a decreased ventricular function in 6 (15%) patients and significant atrioventricular valve regurgitation in 3 (7%). NYHA Class was I in 17 (43%), 11 (2.5%) received cardiac medication, and 8 (2%) had an arrhythmia. Median NT-proBNP was 206 (123-660) ng/ml, VO2max 28±10 ml/kg.
Left ventricle (LV)-mid-CS was higher than RV-mid-CS (-24.9 ±4.1% vs. -20.7±3.7%; p=0.004). LV-GLS was lower than RV-GLS (-15.6±4% vs. -19.9±6.8%; p=0.05). Total Mid-GCS correlated positively with larger EDV (p=0.006), smaller ESV (p=0.001) and EF (p=0.01). Total GLS correlated positively with EF (p=0.003) but not with ventricular volumes. Patients with echocardiographically decreased ventricular function had lower mid-GCS (p=0.03). No correlation was found between strain values and presence of fenestration, postoperative complications, atrioventricular regurgitation, NYHA class, arrhythmia, age at Fontan, hospitalization length, age, body size, interval since Fontan, NT-proBNP, and VO2max.
Conclusions: Myocardial deformation can be assessed with CMR-FT in patients after Fontan palliation. Systemic RV and LV have significantly different strain patterns. Mid-GCS shows a relevant correlation with ventricular volumes and ejection fraction, while GLS correlates only with ejection fraction. No correlation was observed between myocardial deformation and clinical parameters or biomarkers.

MP3-3
Very low progression rate of ascending aorta dilatation in children with isolated bicuspid aortic valve
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Introduction: Bicuspid aortic valve (BAV) is the most common congenital heart disease. Children with BAV have an increased risk of developing aortic valvar disease and progressive ascending aorta dilatation. Studies have linked aortic dilatation to the severity of valvar disease and authors have hypothesized that dilatation is caused by increased shear stress on the aortic wall. Some studies have also reported distinct dilatation patterns according to the type of leaflet fusion. However, we are still unable to predict which children with BAV will present a significantly dilated ascending aorta when entering adulthood.
Objectives: We sough to determine longitudinal risk factors of progressive ascending aorta dilatation in children with BAV and to evaluate the risk of dilatation in patients with isolated BAV (without significant stenosis or regurgitation).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

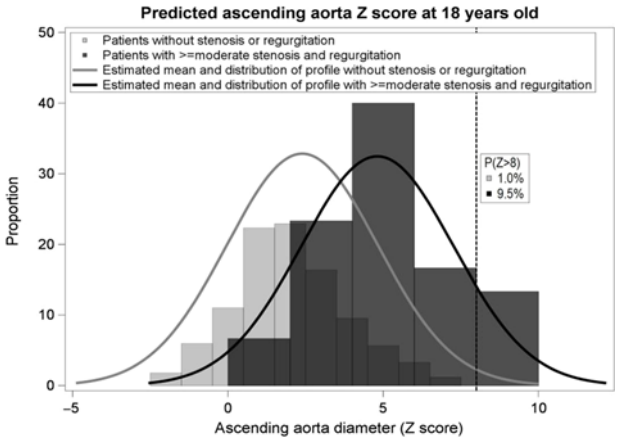

Figure1.

Methods: We extracted all echocardiography reports performed on BAV patients aged <20 years old between 1999 and 2016 at our institution. We collected information on type of fusion, severity of valvar disease, presence of coarctation of the aorta and ascending aorta diameter (normalized to Z-scores). We used multivariate linear mixed models to evaluate the influence of hemodynamic risk factors and BAV type on aortic dilatation rate and then estimated the probability of severe dilatation at 18 years of age according to different hemodynamic profiles.
Results: A total 761 patients (3,148 echocardiograms) were included. Median follow-up was 4.4 [IQR 1.0-8.4] years. Increased aortic dilatation rate was associated with severity of aortic regurgitation and severity of aortic stenosis. In patients with isolated BAV, aortic dilatation rate was low (0.05 Z-score unit per year) and showed no association with fusion type. The estimated probability of developing a dilated ascending aorta with a Z-score > 8 at 18 years of age was 1.0% in patients with isolated BAV and 9.5% in patients with at least moderate stenosis and regurgitation (figure 1). We observed no dissections or surgery for reduction of aortic size during follow-up period.
Conclusions: In children with isolated BAV, the mean rate of aortic dilatation is low but significantly increases in presence of valvar stenosis and regurgitation and is not associated with BAV type.

MP3-4
Maternal obesity and Gestational Diabetes associations with offspring body composition and left ventricular diastolic function in early childhood – RADIEL study follow-up

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Introduction: Maternal obesity and gestational diabetes (GDM) are linked with offspring long-term cardiovascular disease. Fetal programming is hypothesized as an underlying mechanism. We aimed to investigate the influence of maternal adiposity and GDM on child body composition and left ventricular diastolic function in early childhood.

Table 1 Associations with offspring diastolic parameters.

	LAV		S/D		A'	
	r	*p*-value	*r*	*p*-value	*r*	*p*-value
Age	0.16	0.03				
Height	0.42	<0.001				
Weight	0.53	<0.001				
BMI z-score	0.43	<0.001	0.15	0.003	0.22	0.002
Waist-height ratio	0.16	0.02	0.19	0.006	0.27	<0.001
Lean body mass	0.51	<0.001				
Body fat mass	0.46	<0.001				
Body fat percentage	0.25	<0.001	0.21	0.003	0.19	0.007
Maternal height	0.2	0.006				
Maternal lean body mass	0.28	<0.001				

Methods: This observational follow-up study includes 201 mother-child pairs, a subcohort form Finnish Gestational Diabetes Prevention Study (RADIEL). GDM was diagnosed in 96 mothers, 36 of whom required metformin or insulin treatment. Follow-up assessment was performed at 6.1 (+/- 0.5) years postpartum including child echocardiography, child and maternal anthropometrics, body composition, and blood pressure.
Results: Maternal pre-pregnancy body mass index (BMI 30.5 +/- 5.6 kg/m^2) correlated with child BMI z-score (r=0.2; p=0.006; mean z-score 0.45 +/- 0.93 kg/m^2). Left atrial volume (LAV) correlated with child age, body size and composition (Table 1). In a multiple linear regression model LAV was independently associated with child lean body mass and body fat percentage (R2=0.283). Pulmonary vein flow systolic to diastolic ratio (S/D) and mitral annular velocity during atrial filling (A') were associated with child adiposity parameters (Table 1). Left ventricular diastolic parameters including LAV, LAV index z-score, mitral valve E and A waves peak velocities, E/A ratio, S/D, mitral and septal annular velocities (E'and A') were not associated with maternal pre-pregnancy BMI, I trimester glycated hemoglobin (HbA_{1C}), or GDM exposure.
Conclusions: Child diastolic heart function at six years of age is associated with child adiposity. Maternal pre-gestational obesity is reflected in child body composition, increasing long-term cardiovascular risks and implicitly disturbing diastolic heart function. No evidence of fetal cardiovascular programming related to GDM was found in early childhood.

MP3-5
Pronostic factors after arterial switch operation for transposition of the great arteries in children

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Pronostic factors after arterial switch operation for transposition of the great arteries in children
The objective of this study was to assess long-term outcome and factors associated wih mortality in children after arterial switch operation (ASO) for transposition of the great arteries (TGA).
Material and methods: retrospective analysis of all ASO performed from 2000 to 2016 in patients with TGA. Demographics, clinical and biological data, surgical techniques, echocardiographic measurements, and outcomes were collected. Pronostic factors for mortality were assessed.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Results: 364 patients were included (263 males). Antenatal diagnosis was made in 60,4%. Birth weight was 3207,5±511.2g, gestational age was 39.1±1.6weeks. Intramural coronary artery was present in 24 cases (6.6%). Long term follow-up was 9.2±4.8years. Rashkind procedure was performed in 88.7% of the cases. Ventilatory support was needed in 56% and prostaglandins in 85% preoperatively. ASO occurred at the age of 7.44±6.26 days (1 to 56) and 5.6±4.6days after admission. Bypass duration was 123.1±33.3mn. Overall and postoperative hospital stay were respectively 19.9±8.6 and 14.4±7.3days. Survival rates were 95.3% at 1 month, 94.8% at 3, 6 and 12 months and 94.5% at 2.5 years and up to 18 years after surgery. Twenty patients died (5.5%): 17 early postoperatively (4.7%). Time from surgery to death was 64.2±207.9days (0 to 919). Long-term events occurred in 49.6% of the patients: pulmonary stenosis, aortic or mitral regurgitation, residual shunt, coronary lesions (1.7%), arrhythmias (5.5%), cardiac failure. Freedom from late reintervention was 99.7%, 97.4%, 96.2% at 1, 3 and 6 months, 95.9%, 95%, 92.3% and 90.3% at respectively 1, 2, 5 and beyond 8 years after ASO. No Rashkind, postoperative ECMO support, delayed chest closure, bypass duration, troponine level and small birth weight were significant predictive factors of mortality (respectively p= 0.006, p<0.0001, p=0.0007, p<0.0001, p=0.001 and p=0.044). Antenatal diagnosis, gender, gestational age, preoperative ventilatory support, prostaglandin infusion, and SpO2, coronary arteries anatomy or age at surgery were not associated with death.
Conclusion: This study showed that Rashkind procedure may have a favourable impact on prognosis of neonates with TGA. Postoperative ECMO support and troponine level, and small birth weight were associated with worse outcome.

MP3-6
Computational Fluid Dynamics in complex pulmonary artery stenosis

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Introduction: Pulmonary artery (PA) stenosis is common in congenital heart diseases. Understanding flow patterns and hemodynamic parameters is essential to improve diagnosis and treatment for PA stenosis. Studying flow is challenging as current imaging techniques fail to provide a complete overview of all hemodynamic parameters. Computational Fluid Dynamics (CFD) is a tool mainly used in (medical) engineering to study flow characteristics. It enables visualization of complex flows and provides detailed information on flow velocities, wall shear stress (WSS), energy losses and turbulence. Our objective was to create a CFD model for patient specific flow evaluation in complex PA stenosis.
Methods: A 3D patient specific model was created for 4 cases. The PA bifurcation was isolated by manual segmentation of DICOM files obtained by 3DRA during catheterization. The model was manually smoothened, peripheral pulmonary arteries were removed and a mesh was generated. Individual transient mass flow inlet and pressure outlet boundary conditions were defined. Cases were solved and post-processed using ANSYS Fluent (ANSYS Inc, Canonsburg, Pa). Convergence was accepted at residuals <1e-4.
Results: All cases showed flow acceleration over the stenotic areas. WSS was unequal distributed and there was major energy loss over the stenosis. Turbulence was seen in areas with low flow velocity and in vessels with post-stenosis dilation. Velocity and pressure outcomes for each case were comparable with echographic and catheterization data. Time from pre-processing to post-processing varied between 10 and 13 hours.

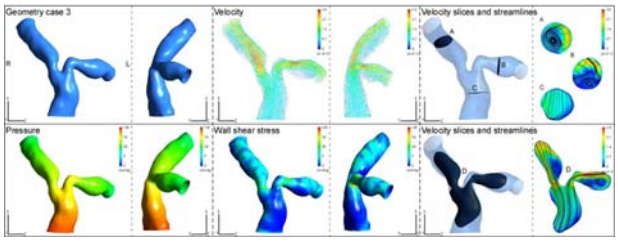

Figure 1.
CFD analysis of the right (A), left (B) and main (C) PA of a patient with severe stenosis. Velocity, pressure and WSS outcomes are shown. The right images show flow patterns and velocity contours on slices through A, B, C and D.

Conclusions: Patient specific CFD analysis creates a better understanding of hemodynamic parameters and flow characteristics in complex PA anatomy. This allows for evaluation of different treatment options and may provide insight in mechanisms causing re-stenosis after PA stenting. The used method generates realistic outcomes which are comparable to values obtained by other imaging techniques. In order to further improve the model stent simulation and compliance should be considered in future analysis.

MP3-7
Liver stiffness: a useful tool in the longitudinal follow-up of patients with Fontan circulation

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Introduction: Congestive hepatopathy usually appears and develops after a Fontan operation (FO), often without obvious clinical features, but it may lead to life-threatening complications.
Objective: We aim to assess the potential usefulness of the liver stiffness (LS), assessed by elastography, in the longitudinal follow up of Fontan patients.
Methods: In our center, patients with a Fontan circulation were prospectively evaluated since 2012 through an annual work up including physical examination, laboratory tests, trans-thoracic echocardiography and LS using transient elastography (TE). This work up was also performed in case of clinical complications, which were classified as follows: cardiac complications (including arrhythmias or catheter interventions) and subdiaphragmatic complications (including clinical sign of portal hypertension or protein losing enteropathy (PLE)).
Results: Fourty eight patients (21.7± 8.2 years of age and 9.7±6.5 years post-Fontan) were included, 28 of them (58%) had least two LS measurement. Mean time between first and last LS measurements was 3.27 ±1.9 years. Mean LS at baseline was 15.3±6.9kPa (4.3-47.2 kPa) No significant correlation was found between LS and age (r=0.8, p=0.73), time since Fontan surgery (r=0.3, p=0.64). LS did not vary regarding the presence of a fenestration (15±6.8 vs 15.1±6.7 kPa, p=0.82).
During the follow-up, a clinical complication occurred in 19 patients (39.6%) including 8 cardiac complications and 11

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

subdiaphragmatic. Among the cardiac complications group, 6 had atrial flutter and 2 had catheter-based intervention (occlusion of vein-venous collateral). Among the subdiaphragmatic complications group, 3 had PLE and were referred for heart transplant and 8 developed portal hypertension. LS was significantly higher in patient with liver complication (17.2±7.7 vs 13.8±5.9, p<0.01). Patient with an increasing LS value during the follow up had a higher complications rate than patients with a decreasing or unchanged LS (4/11 (36%) vs n=5/17 (29%); p<0.04).
Conclusion: LS measurement using TE is a good tool for the non-invasive follow-up of patient palliated with FO. Indeed, a significate elevation of the LS is associated with the occurrence of liver and/or cardiac complications.

MP3-8

The predictive value of cardiopulmonary exercise testing in children with congenital heart disease

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Background: Children with congenital heart disease (CHD) are at increased risk to develop late complications, e.g. arrhythmias, residual valvar dysfunction and heart failure, possibly leading to re-intervention and/or rehospitalization. In clinical practice, predicting which patients will develop complications remains difficult. In adults with CHD, cardio pulmonary exercise test (CPET) has been used to identify those at risk for morbidity and mortality, but its value in children with CHD has not been established. Our aim was to investigate the predictive value of CPET for morbidity and mortality in children with CHD.
Methods: We retrospectively assessed patients with CHD who performed a CPET between 2001 and 2017. We performed clinical follow up, starting at the day of CPET. We excluded patients who underwent an intervention within 3 months after the CPET. Clinical data were extracted from the hospital medical records. Primary endpoints were mortality or heart transplantation and a composite endpoint (CE) of cardiac hospitalization, arrhythmia, cardiac surgery, percutaneous intervention or the use of heart failure related medication. Patients who underwent an intervention within 3 months after the CPET were excluded from the analyses. Unpaired t-tests, univariate and multivariate Cox regression (backward stepwise selection) were used for analyses.
Results: 402 children with CHD were included in this study. Six patients died during an mean 6.7 +- 3,5 years follow-up. The absolute number of deaths did not allow regression modelling. A total of 135 patients reached the composite endpoint. Univariate analyses revealed nine patient characteristics and three CPET parameters as significant predictors of the composite endpoint. After multivariate analysis, peakVO2/kg was the only CPET variable in the final model; the other parameters were number of thoracotomies, use of β-blockers, and valvular inflow/outflow dysfunction (see figure). Cyanosis and load/kg lost statistical significance after correcting for multiple testing. Stratification on univentricular vs. biventricular circulations showed no essential differences.
Conclusion: In a paediatric cohort of patients with congenital heart disease, PeakVO2/kg predicted a composite cardiovascular endpoint independently of patient characteristics. These findings can be used to develop a prediction model for children with CHD.

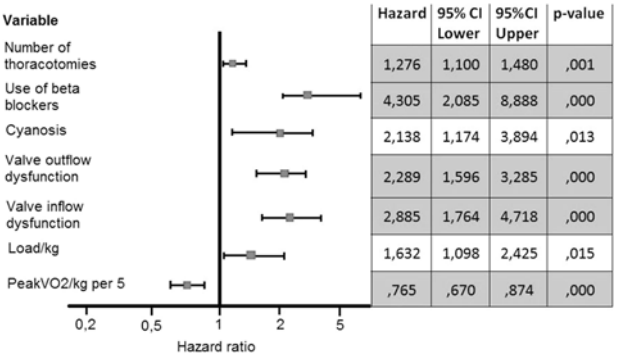

Figure.
Hazard ratios with confidence intervals in multivariate analysis (backward stepwise selection).

MP3-9

Contrast Echocardiography for Hepatopulmonary Syndrome Risk Stratification in Children With Normal Arterial Oxygenation

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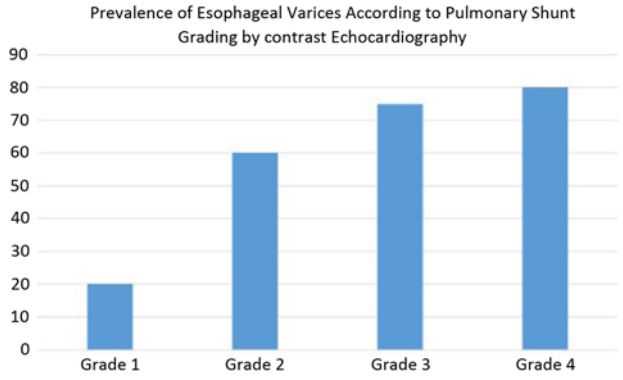
Background: Hepatopulmonary syndrome (HPS).is defined as a triad including liver disease (and/or portal hypertension), intrapulmonary vascular dilatations (IPVD) and abnormal arterial oxygenation, and is associated with a pretransplantation mortality of 25%-46% in children. However, in the early phases of the disease abnormal oxygen saturation might not be found and thus echocardiographic identification of IPVD by bubble study contrast echocardiography (CE) is of paramount importance for risk stratification. Aim of this study was to determine the prevalence and associated signs and symptoms of IPVD (diagnosed by CE) in children with liver disease and portal hypertension without HPS.
Methods: The study population included 44 patients with liver disease and portal hypertension with $SpO_2 \geq 99\%$ (20 girls; age 12±4 years, range 2-18). IPVD were diagnosed by using CE and they were based on the presence of microbubbles in the left atrium with >3 cycles and they were graded according to the Barzilai score (grade I to IV). Patients with evidence of intracardiac shunt were excluded (n=3).
Results: Specific diagnoses for the patients included: portal vein cavernoma (58%), biliary atresia (27%), congenital hepatic fibrosis (10%) and others (5%). Among the 41 included patients, only 10 patients showed no evidence of pulmonary shunting, while a positive CE was found in 31 patients (or 76%). In details: Grade I was found in 14 (34%) patients, Grade II in 8 (20%), Grade III in 4 (10%) and Grade IV in 5 (12%). A significant association was found between the severity of the disease, according to the semi-quantitative Barzilai score and the total serum proteins (r= -0.325) and INR values (r=0.261; both p<0.05). No association was found between the length of the disease and the anthropometric parameters (weight, length, BMI, Z score BMI) with the presence of a shunt. A comparison of the groups, according to the Barzilai score at the CE, demonstrated a significant association with the presence of the esophageal varices (p for trend= 0.042; Figure).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



Conclusions: Prevalence of positive CE in children with liver disease is a common finding. Accurate grading of positive CE is of paramount importance, as despite normal arterial oxygenation in the presence of significant shunting (Barzilai grade>II), signs of disease severity can be found, including abnormal blood coagulation associated with the presence of significant esophageal varices, thus identifying pediatric patients at high risk of HPS development.

MP3-10
Does diastolic dysfunction improve following the Ross procedure?

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Introduction: Aortic valve stenosis causes left ventricular hypertrophy (LVH) and myocardial fibrosis resulting in diastolic dysfunction. Advanced diastolic dysfunction at baseline prior to aortic valve replacement has been associated with increased mortality and morbidity. Reverse LV remodelling and resolution of diastolic dysfunction following aortic valve replacement takes two to six years in adults. Little is known about the reversibility of diastolic dysfunction in children. We sought to determine if diastolic function improves in children with aortic valve disease following the Ross procedure compared to healthy controls. Methods: Retrospective study of patients with aortic valve disease following the Ross procedure, healthy controls were also included (n=92). Echocardiograms prior to and at 1 year following the Ross procedure were reviewed. Systolic function was assessed by shortening fraction (SF). LV mass was recorded and indexed to height$^{2.7}$ (LVmassHt). Diastolic measures included: mitral valve velocities (E, A, E/A ratio, E/E'), E wave deceleration time, and TDI E' and A' in the lateral LV (LatLV), IVS, and right ventricular (RV) walls. Medians and interquartile ranges are reported. A Wilcoxon rank sum test was done to compare patients pre vs post Ross procedure, and post Ross procedure vs controls. A p-value of <0.05 was considered statistically significant.
Results: Forty three patients (31 male) were included with follow up data available on 31 patients. Age at surgery was 9 years (4-14 years). LV SF was normal in patients' pre Ross [38% (34-44%)]. At 1 year, LVmassHt was lower compared to pre Ross values (59 vs 76 gm/ht$^{2.7}$; p<0.001), however was not normal compared to controls (59 vs 39 gm/ht$^{2.7}$; p<0.001). There was no change in LV diastolic function between pre and post Ross procedure, and diastolic function was significantly lower in post Ross patients than controls (Table 1). Conclusions: Diastolic dysfunction persists in pediatric patients with aortic valve disease at 1 year following the Ross procedure. Follow up over time will determine whether diastolic dysfunction returns to normal.

		Pre Ross	Post Ross	p value[a]	Controls	p value[b]
Mitral inflow velocity	E (m/s)	1.15 (0.90-1.30)	1.10 (0.90-1.3)	NS	0.85 (0.80-1.0)	<0.001
	A (m/s)	0.5 (0.4-0.8)	0.6 (0.5-0.8)	NS	0.4 (0.3-0.5)	<0.001
	E/A ratio	2.0 (1.3-2.9)	1.7 (1.4-2.2)	NS	2.4 (2.0-2.6)	<0.001
	Decel time (ms)	155 (138-180)	171 (148-207)	NS	135 (115-154)	<0.001
	E/E'	7.4 (6.3-10.2)	7.0 (5.7-9.2)	NS	5.3 (4.5-6.4)	0.001
TDI LatLV	E' (cm/s)	13 (12-16)	14 (12-17)	NS	16 (14-18)	0.044
	A' (cm/s)	5 (4-6)	5 (4-6)	NS	6 (5-7)	0.013
TDI IVS	E' (cm/s)	13 (12-16)	14 (12-17)	NS	16 (14-18)	<0.001
	A' (cm/s)	5 (4-6)	5 (4-6)	NS	6 (5-7)	NS
TDI RV	E' (cm/s)	13 (12-16)	14 (12-17)	NS	16 (14-18)	<0.001
	A' (cm/s)	5 (4-6)	5 (4-6)	NS	6 (5-7)	<0.001

MP3-11
Gut inflammation in Fontan patients is associated with increased enteric protein-loss, augmented systemic inflammation and alterations in vitamin D homeostasis

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Objectives: Gut inflammation (GI) has been observed in Fontan patients with protein-losing enteropathy (PLE). The clinical relevance of this finding is unknown. The aim of the present study was to identify factors associated with IF in a Fontan cohort.
Methods: A retrospective chart review was performed of Fontan patients who had been screened for both enteric protein-loss and presence of GI, by measuring fecal alpha-1 antitrypsin (A1AT) and fecal calprotectin (FC) levels, respectively. Associations between laboratory parameters (serum albumin level, markers of systemic inflammation, vitamin D metabolism) and clinical characteristics were explored. Patients without ≥ moderate ventricular dysfunction, ≥ moderate valvular regurgitation, cyanosis, or PLE, and classified as NYHA class I were defined as good Fontan. Patients not fulfilling these criteria were classified as failing Fontan.
Results: From 2011 to 2018, 41 Fontan patients (31.7% female, age 9.3 ± 3.6; PLE: n = 18, 43.9%) were screened. Increased FC levels (> 50 ug/g) were found in 16 patients (39%, PLE n = 10). A strong correlation between FC and A1AT levels was found (r 0.689, p <0.0001). This association was independent of having a good Fontan circulation, presence of GI or PLE (all p < 0.05).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

GI was found in 6 Fontan patients without PLE (14.6%). Interestingly, significant enteric protein-loss developed in 4 of these patients (median A1AT 683 ug/g, normal 100 – 500 ug/g) within 11 - 26 months. Furthermore, PLE patients with active GI had lower albumin levels, lower lymphocyte count, higher NLR and A1AT than PLE patients with normal FC concentrations (all $p < 0.05$). Furthermore, strong correlations were found between FC, measures of systemic inflammation, serum albumin levels, and markers of vitamin D metabolism in PLE patients, patients with GI, and failing Fontan patients (all $p < 0.05$), but not in good Fontan patients (all $p > 0.1$).
Conclusions: GI seems an emerging mechanism of disease in Fontan, and is strongly associated with severity of enteric-protein loss, augmented systemic inflammation, and altered vitamin D metabolism. Future studies are needed to determine whether alterations in intestinal function are responsible for these findings.

MP3-12
Functional Outcome in Children and Adolescents with Coarctation of the Aorta
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Introduction: Coarctation of the Aorta (CoA) was assumed being one of the congenital heart defects not associated with major long-term problems. Meanwhile it is known that there is an association to some side defects like vascular dysplasia which still exists after surgical correction. Due to this CoA patients are at higher risk for long-term vascular impairments and higher cardiac morbidity. This study investigates functional outcome measures in children with CoA in comparison with a healthy control group (CG).
Methods: From May 2014 to October 2018, we examined 75 children (42.7% girls, 12.9 ± 3.6 years) with CoA for their arterial stiffness, Intima-Media Thickness (IMT) and Health-Related Physical Fitness (HRPF). The functional arterial stiffness measures, central systolic blood pressure and pulse wave velocity (PWV) were analyzed with an oscillometric device. Structural changes were measured by ultrasound of the arteria carotis communis IMT; in addition HRPF was tested by five tasks of the FITNESSGRAM®. The CG consisted of 2002 children (48.9% girls, 12.8 ± 2.8 years) they were recruited within two recent school projects.
Results: In comparison to the CG, adjusted for age and sex, the CoA patients showed worse functional arterial stiffness measures with a 2.9mmHg higher central systolic blood pressure (CoA: 104.0 ± 7.7mmHg vs CG: 101.1 ± 7.7mmHg; $p=0.007$) and a 1.8mmHg lower peripheral diastolic blood pressure (CoA: 64.5 ± 7.2mmHg vs CG: 66.3 ± 7.2mmHg; $p=0.049$), whereas no significant difference in peripheral systolic blood pressure. Structural changes were also present in CoA patients with a 0.028mm thicker intima media vessel wall (CoA: 0.481 ± 0.037mm vs CG: 0.454 ± 0.034mm; $p<0.001$). Compared to the CG and adjusted for age and sex, children with CoA presented significantly lower HRPF (z-score -0.47 ± 0.65; $p<0.001$; 31th percentile).
Conclusions: Children with CoA show significant impairments in all parts. Especially functional arterial stiffness and IMT measures for structural changes are in close relation with higher risk for severe vascular and cardiac morbidities. Based on these aspects it is important to ensure a comprehensive and structured aftercare monitoring to prevent cardiovascular morbidity as soon as possible.

MP4-1
Early Detection of Sudden Infant Death Syndrome: Electrocardiogram, Genetic Analysis and Familial Assessment
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Background: Arrhythmogenic diseases have been proven to cause infant sudden death. Some of them can be diagnosed performing a 12-lead ECG at birth.
Objective: We pretend perform an ECG in all births in order to identify the percentage of any malignant electrical alteration in neonatal population.
Methods: We have performed a prospective analysis of ECG performed at 24 to 72 hours of life to new-borns of a single centre. Of those which QT interval was above normal, saliva was collected for DNA genetic analysis using a gene panel related to sudden death using ultrasequencing technology. Clinical and genetic analysis of the family members has been performed accordingly.
Results: During the first year of the project we have performed 600 ECG in new-borns (51% males). We have identified 14 cases of prolongation of the QTc above 470ms and ECGs of the first-degree family members has been performed. In 7 of them (QTc below 490ms) normalization of the ECG has been observed along the first 6 months of life and no family members have been detected abnormal. In the remaining 7 cases with persistent QTc interval above 490 ms., a genetic analysis has been performed. In 6, family members ECG were abnormal. In 5, a rare genetic variant has been described (one of them, de novo).
Conclusions: Neonatal ECG screening allows early detection of potentially lethal cardiac diseases. Genetic study helps identifying the underlying mechanism of the disease. Family cascade study is mandatory in order to detect other disease carriers.

MP4-2
Hybrid therapy of anti-tachycardia pacing and catheter ablation of atrial tachycardia for patients with complex congenital heart disease
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Background: Device therapy of bradycardia may be required for patients with congenital heart disease (CHD) after surgery. Additionally, such patients may have complicated substrates of atrial tachycardia (AT) resistant to antiarrhythmic drugs and catheter ablation (CA).
Objectives: The aim of this study is to evaluate the clinical outcomes of hybrid therapy with anti-tachycardia pacing (ATP) and CA for AT in patients with complex CHD.
Methods: Twenty-four CHD patients after cardiac surgery (two-ventricular repair in 6, Fontan palliation in 12, atrial switch operation in 4, and palliative procedure in 2) with AT and pacemaker-indicated bradycardia were investigated retrospectively.
A pacemaker was implanted in 23 patients for sick sinus syndrome in 18 and atrioventricular block in 5 and an implantable cardioverter defibrillator in 1 patient for ventricular tachycardia at

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

the age of 9-59 years old. CA was performed in all patients and the acute success (no inducible AT) rate was 67% (16/24).
Results: Appropriate ATP and successful termination of AT was observed in 38% (9/24) during the average follow-up period of 4.0 years after implantation.
However, ATP did not start in 25% (6/24) because of slow AT in 4 patients and of 1:1 atrioventricular conduction during AT in 2 patients.
Conclusion: Although CA was not always successful for complex CHD, an automatic atrial ATP option showed effective additional treatment for refractory AT. Further blush-up of the ATP programming should be required.

MP4-3

S-IcD registry in European paediatriC and Adult patients with congenital heaRt defects: preliminary results of the SIDECAR project

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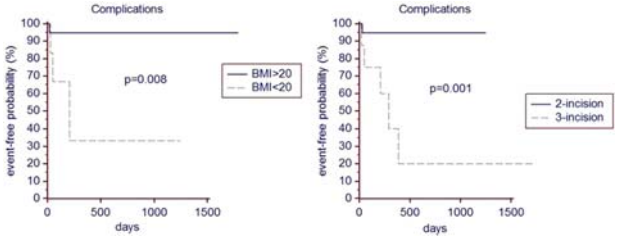
Introduction: Use of the subcutaneous implantable cardioverter-defibrillator (S-ICD) to prevent sudden cardiac death is increasing. Few data exist on S-ICD in young patients. We report preliminary data from a multicenter European registry of paediatric and young adult patients who underwent S-ICD implantation
Methods: Observational, prospective, non-randomized, standard-of-care study on S-ICD implantation/follow-up in young patients with inherited arrhythmias (IA), cardiomyopathies, and congenital heart defects (CHD). 27 patients (11 CHD, 14 Cardiomyopathies, and 2 IA), mean age 17± 6 years, 11 of them <18 years, with body mass index (BMI) 23.5 ± 4.5, underwent S-ICD implantation (primary prevention 69%). The first 8 patients underwent a standard implantation procedure (three surgical incisions), the following 19 (70%) a two-incision procedure.
Results: No intraoperative complications occurred. Over the 17 months median follow-up (25th–75th percentiles, 5–35) 3 patients (11%) received appropriate and 2 (7%) inappropriate shocks. Four patients (15%) had device-related complications requiring surgical intervention: three skin erosions at the superior parasternal incision, one pocket infection. A higher risk of complications was seen in patients who underwent standard procedures [hazard ratio (HR) 14.7, 95% confidence interval (CI) 2.34 to 93.03;P = 0.001] and those with BMI <20 (HR 11.06, 95% CI 1.01–121.07; P = 0.008).
Conclusions: These preliminary results of a multicenter European paediatric registry suggest that S-ICD is safe and effective with low rates of inappropriate shocks. Improvement of implantation techniques seems associated with better outcome



MP4-4

Electrophysiologic characteristics and catheter ablation results of tachycardia-induced cardiomyopathy in children with structurally normal heart

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Objectives: Tachycardia induced cardiomyopathy (TIC) is a rare but curable form of cardiomyopathy. For this reason, an early diagnosis is crucial. The data about TIC ablation in childhood are limited. The aim of this study is to present electrophysiologic characteristics and catheter ablation results of TIC in children with structurally normal heart.
Patients and Methods: We reviewed retrospectively records of 945 transcatheter electrophysiological study(EPS)-ablation procedures done in our clinic between November 2013-November 2018, and found a total of 25(10 females, 40%) patients who underwent EPS and ablation due to TIC. The EnSite™3-D mapping system (St.Jude Medical Inc.,St. Paul,MN,USA) was used in all patients.
Results: The mean age of the patients was 6,96±5,92 years (median:5,84 and range: 2 months-17,8 years) and the mean body weight was 27.96±23,63 (median:22 kg, range: 2.5-85) kg respectively. Tachycardia substrates were as follows; focal atrial tachycardia(n=8), concealed accessory pathways(n=4), permanent junctional reciprocating tachycardia(n=4), Wolff-Parkinson-White preexitation/syndrome (n=3, one patient intermittant), atypical atrioventricular nodal reentry tachycardia(aAVNRT;n=2), ventricular arrhythmia(n=2), multifocal atrial tachycardia + atrial flutter (n=1), Mahaim Tachycardia (n=1). Successful ablation was performed in 23/25 patients (26/28 substrate), and 10 cryoablations and 15 RF ablations were used. Mean procedural time was 1688 ±57,8 (median:170, range: 72-310) minutes and flouroscopy was used in 19/25 patients(%76) with a mean duration of 7,83±9,94 (median:3,57,range:0-39.4). During a median follow-up of 22.2 ±13.55 months (median:17; range:4-54), tachycardia recurrence was observed in 2 patients, which had successful repeated RF ablation procedure. Overall success rate was 23/25 patients (92%) and 26/28 (%93) substrates. There were three complications overall;one transient pericardial effusion (during transseptal punction in a 12 years old FAT patient), and one permanent incomplete RBBB (in a patient 3 months old, with a right anteroseptal/parahisian AP), and the last one transient ST elevation during ablation (in an 11 years old patient with FAT, L-UPV). Average left ventricular EF and FS values of patients before the ablation were 38.8±7.1%(26-48%) and 23.3± 4.8%(12-30%) respectively. In a mean period of two months later, the mean EF and FS values reached 53.6±12.7% (35-70%) and 32.4±4.9%(19-40%) respectively.
Conclusion: Transcatheter ablation is an effective and safe method for the treatment of TIC, even in small children and infants.

MP4-5

The results of thoracoscopic procedures in pediatric patients with life-threatening cardiac arrhythmias and conduction disorders

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The objectives: To evaluate the outcomes of videoassisted thoracoscopic surgery (VATS) as an adjunctive therapeutic approach in

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

pediatric patients (pts) with cardiac arrhythmias and conduction disorders.
Methods: In 2016-2018 yrs, 22 pts aged 4 to 15 underwent VATS at our institution. Group I (n=16) – pts with long QT syndrome (LQTS) and catecholaminergic polymorphic ventricular tachycardia. Group II (n=6) – pts (body mass less than 15 kg) with third-degree atrioventricular block (n=5) and Sinus node dysfunction (n=1). In Group I all pts underwent left cardiac sympathetic denervation (LCSD) (low pol of Th1Th4), 8 pts (50%) received implantable cardioverter-defibrillators (ICDs) before or during LCSD. In all cases betablockers therapy were not effective before LCSD. Invasive electrophysiological study (EPS) was performed before and after sympathetic denervation. In pts with ICDs EPS was performed via device. In Group II permanent cardiac VVIR pacemaker (PM) with bipolar leads (2 pts) and unipolar screw-in leads (4 pts) were implanted using VATS. Followup period was 3 - 24 months with continuous beta-blockers therapy.
Results: The trend to increasing of right ventricular effective refractory period have been found just after LCSD. We observed no cardiac arrhytmias in 15 pts (93.75%) in Group I. Nonsustain ventricular tachycardia in this group was induced during exercising test in 1 child (6.25%). Pacing and sensing paraments were appropriate in Group II. There were no any complications in bouth groups.
Conclusion: LCSD is a minimally invasive technique that results in good benefits in patients with LQTS. Permanent PM implantation using VATS could be selected for pediatric pts with low body mass. VATS was confirmed to be an effective adjunctive therapy in pediatric pts with cardiac arrhythmias and conduction disorders.

MP4-6

Revealing cardiac microstructure in a human fetal heart of 8 weeks of gestation with synchrotron-based X-ray phase contrast tomographic imaging

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Introduction: Understanding the complexity of heart morphogenesis and the associated functional consequences of congenital heart disease is essential for providing appropriate treatment strategies. Since our knowledge on the microstructure of the whole fetal & paediatric heart is limited, novel imaging approaches offered by synchrotron facilities can provide structural detail currently not available otherwise. Our aim is to visualise and quantify cardiac microstructure in fetal hearts at different stages of development using synchrotron-based X-ray Phase-Contrast tomography Imaging (X-PCI).
Methods: A normal fetal heart of 8 weeks and 6 days of gestation was selected from the from the Ospedale Maggiore Policlinico (Milan, Italy). While the specimen was fixed in formalin, it was placed in water as supporting medium for acquisition. X-PCI was performed at 1.625μm resolution at the TOMCAT Beamline (Swiss Light Source, Paul Scherer Institut, Villigen, Switzerland) using an energy of 20 keV. Several acquisitions were necessary to cover the whole heart along its long axis. The image series were reconstructed using Gridrec algorithm. Orientation of myocytes aggregates was computed using an in-house structure tensor algorithm.
Results: Fig.1(a) Two images of the gross specimen with scale - base to apex 2mm. Fig.1(b)-(c) show longitudinal (4-chamber) and

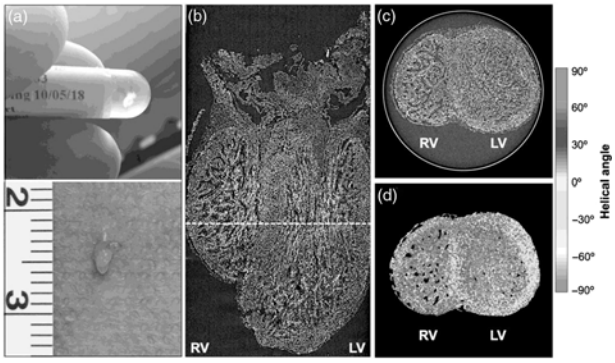

Figure 1.

short axis maximum intensity projection slices, respectively, from the X-PCI image dataset showing detail of myocardial structure. The ventricular myocardium is composed mainly of trabeculations while the compact myocardium is thin and under-developed. Taking both trabecular and compact myocardium together there is organisation even at this early gestation (see Fig.1(d)) with a clear change in helical angle (from 60º to -60º) from endo to epicardium, especially the septal wall.
Conclusions: We managed for the first time to image a normal fetal heart with high-resolution and in 3D at an early stage of development, resolving detail of myocyte aggregates and providing information on cardiac microstructure without the need for sample processing or sectioning.

MP4-7

Prenatal diagnosis improves pre-operative status and outcomes after neonatal cardiac surgery: Impact of a Fetal cardiology program from a lower middle-income country

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Introduction: Congenital heart defects (CHD) are a major contributor of infant mortality in many parts of the world. Prenatal diagnosis of CHD has become standard of care in most high-income countries; however, in low and middle-income countries (LMICs), this is still an unexplored concept. The Fetal Cardiology division was developed at the Amrita Institute of Medical Sciences and Research Centre (AIMS), Kerala in 2008 with the goal of improving peri-operative outcomes for neonates with critical CHDs through availability of prenatal diagnosis and planned peri-partum care.
Methods: Specific interventions performed included establishment of a comprehensive multidisciplinary fetal cardiac team, creation of a fetal cardiac database to track outcomes and a state-wide prenatal CHD screening capacity building program for obstetricians and radiologists with development of a referral pathway and network. This was done in collaboration with National Health Mission, Kerala, and an international non-governmental organization (Children's HeartLink). Trends in referral patterns of fetuses diagnosed with CHD, planned deliveries, pre-operative cardiac status, operative outcomes and costs were analyzed and compared with neonates diagnosed with CHD after birth for the period 2008-2017.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Results: Our interventions created a culture of peri-natal cardiac care which resulted in:

- A three-fold increase (66 to 198) in number of referrals of neonates with diagnosis of fetal CHD.
- A 4-fold increase (9% to 38%) in the proportion of neonatal cardiac admissions with prenatal diagnosis between 2008 and 2017.
- A significantly better pre-operative status in prenatal group (median cardiac score 1 vs 3; p < 0.001; median Ca-TRIPS score 6 vs 8; p 0.001).
- Prenatal group underwent surgery earlier (7.5 + 6.5 vs 10.5 + 8 days; p=0.02)
- A three-fold reduction in surgical mortality in prenatal group (2.9% vs 9%; p =0.04)
- Lower out of pocket expenses for patient families in prenatal group (median INR 63500 vs INR 134,000; p 0.02)

Conclusions: Through concerted efforts, we were successful in establishing a state-wide network for prenatal diagnosis of CHD in Kerala, resulting in significantly improved peri-operative outcomes of neonates with critical CHD with substantial reductions in costs of care, thus making it an optimal strategy for pediatric cardiac care in LMICs.

MP4-8
Parental needs and factors affecting counseling success for prenatal congenital heart disease

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Objectives: To explore parental needs and affecting factors for counseling success after prenatal diagnosis of congenital heart disease (CHD).
Methods: Counseling success after prenatal diagnosis of CHD was measured in five dimensions (Transfer of Medical Knowledge; Trust in Medical Staff; Transparency Regarding the Treatment Process; Coping Resources; Perceived Situational Control) by a pretested standardized questionnaire.
Likert scaled and open-ended questions are combined with sociodemographic data.
Data analyses were conducted with regard to influencing factors and correlations by IBM SPSS® V. 25.
Results: 59 individuals (n=38 female, n=21 male) were interviewed in a tertiary medical care center.
Gestational age at diagnosis and first parental counseling was 28+5 weeks (median).
Parental counseling was performed overall 4 times (median), mainly by pediatric cardiologists (85%) and/or MFM specialists.
Overall counseling was successful in 45.3% and satisfying in 52.8%; 1.9% of parents were dissatisfied.
Counseling was less successful for the dimensions "Transfer of Medical Knowledge" and "Perceived Situational Control". Success rates were higher if additional written information or adequate web sources were provided.
Length of consultation was positively correlated to counseling success for the dimensions "Transfer of Medical Knowledge" (r=0,278*), "Transparency Regarding the Treatment Process" (r=0,330*), and "Perceived Situational Control" (r=0,210). However, the absolute number of parental consultations had no effect on counseling success in any dimension.
Interruptions during consultations were negatively correlated to the dimensions "Trust in Medical Staff" (r=-0,258*) and "Transparency Regarding the Treatment Process" (r=-0,203).
If cardiac diagnosis was graded as high-risk CHD overall counseling success was lower.
The presence of chromosomal abnormalities (n=7) had no effect on any dimension.
If parental mother tongue was different from the language the counseling was conducted, success rates were lower for "Transfer of Medical Knowledge", "Trust in Medical Staff" and "Coping Resources".
Conclusions: These data indicate that parents after fetal diagnosis of CHD need uninterrupted counseling of adequate duration. In addition, written information or links to adequate web sources seem necessary, ideally in the parental mother tongue. High-risk CHD needs more attention for counseling.
Further analyses of sociodemographic factors are necessary to identify parental needs for counseling. The mid-term goal is to propose evidence-based guidelines.

MP4-9
First-degree relatives of patients with hypoplastic left heart syndrome are at higher risk for cardiac malformation

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Introduction: Hypoplastic left heart syndrome (HLHS) is a rare and severe congenital heart defect characterized by a variable degree of underdevelopment of the left-sided heart structures. A higher incidence of cardiovascular malformations (CVM) in relatives of HLHS-patients and a familial clustering of HLHS has been suggested in previous studies. However these assumptions are mostly based on epidemiologic studies, detailed echocardiographic screening in affected families to detect subclinical malformations such as bicuspid aortic valves, is rare.
Objective: This study aims to investigate patterns of familial aggregation of cardiovascular malformations in families affected with hypoplastic left heart syndrome.
Methods: First-degree relatives of HLHS patients (to date n=38) were prospectively recruited at our institution. Clinical examination, 2-D and Doppler transthoracic echocardiography were performed as well as a detailed review of the medical records.
Results: We investigated the families of 21 HLHS-patients. In three families more than one child was born with a congenital heart defect (CHD). One sibling of an HLHS-patient was also affected by HLHS, one suffered from tetralogy of Fallot and the third war born with a ventricular septal defect. Transthoracic echocardiography was performed in first-degree relatives (n=35). A cardiac malformation was found in four participants (11,4%). Two of them had a bicuspid aortic valves (BAV), one severely dysplastic with moderate aortic regurgitation. One father was born with an atrial septal defect with suspected partial anomalous pulmonary venous drainage (PAPVD). One mother was suffering from dilatative

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

cardiomyopathy after an episode of myocarditis. Of the examinated relatives one did not have previous knowledge about the CVM. Overall the incidence of CVM was 18,4% compared to 1% of the average population.
Conclusion: The incidence of cardiovascular malformations was highly increased amongst first-degree relatives of HLHS-patients compared to the average population. Therefore establishing a screening program for congenital heart disease should be considered in families affected with HLHS to prevent further morbidity.

MP4-10

Antenatal Detection of d-Transposition of the Great Arteries (d-TGA) and Impact of Standardised Fetal Heart Screening in Queensland Over A Ten Year Period

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Aims: Dextro-Transposition of the Great Arteries (d-TGA) is known to be one of the more difficult diagnoses to make on antenatal ultrasound screening. We aimed to examine the rates of antenatal diagnosis of d-TGA overall in Queensland, and more specifically in metropolitan Brisbane versus regional Queensland over a ten year period and to assess the impact of targeted antenatal screening education on these rates.
Methods: Data was collected retrospectively from hospital records. All patients diagnosed with d-TGA either antenatally or postnatally in Queensland between July 2008 to December 2017 were included. The patients were divided into two cohorts to assess antenatal detection rates in the pre- and post- education eras (2008-2011 versus 2012-2018), and into metropolitan versus regional groups to examine the impact on antenatal detection rates in the different settings.
Results: 128 neonates were identified with a diagnosis of d-TGA. The overall antenatal detection rate from July 2008 to December 2017 was 61.7%. From 2008 to 2011 (n=47) the overall detection rate was 48.9%, compared to 69.1% in the 2012 to 2018 group (n=81). When analysed as regional Queensland (n=73) versus metropolitan (n=55) groups across the two time periods, the regional Queensland detection rate increased from 35.7% to 59.1%, while the metropolitan detection rate increased from 68.4% to 81.1%.
Conclusions: Rates of antenatal diagnosis of d-TGA in Queensland compare favourably with internationally published rates, although there remains difficulty in consistently diagnosing this lesion. Targeted education of sonographers performing antenatally screening, in particular in regional areas, appears to contribute to an increase in diagnostic rates.

MP4-11

Contemporary study of fetal right aortic arch: change in detection rate, status of associated anomalies and perinatal outcomes

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Objective: To evaluate the prenatal findings of right aortic arch (RAA), its associated anomalies, and the outcome.
Methods: We reviewed all fetuses with RAA diagnosed between 2010 and 2018 in two centres, from the University Hospital of Wales and Kocaeli University Hospital. Prenatal findings of all fetuses with aortic arch anomalies, intracardiac, extracardiac and genetic abnormalities were studied.
Results: A right aortic arch was identified in 52 fetuses. Mean gestational age at diagnosis was 23 weeks (range, 19-33 weeks). There were 23 cases of isolated aortic arch. Associations with additional intracardiac malformations were found in 29 cases: Tetralogy of Fallot in 10, heterotaxy syndromes in four, double outlet right ventricle in four, VSD in four, crossed pulmonary artery in two and others in five fetuses. The detection rate of RAA increased over the study period from occasional encounter to frequent findings after adopting a three-vessels and trachea view in the screening planes. However, in the three quarters of the study period the majority of patients were referred for a suspicion of congenital heart disease on obstetric scanning. The RAA was suspected with the aid of the three-vessels and trachea view in 24 fetuses by obstetricians or sonographers. The RAA was more frequently detected by non-paediatric/fetal cardiologist in isolated cases (16/23) then the RAA with cardiac abnormalities (8/26).). The karyotype and 22q11 status were checked in 22/52 cases: six had confirmed chromosomal anomalies, two 22q11, three trisomy 18, and one 46XX inv-(9)-(p11q12)(20). The rates of chromosomal abnormalities and 22q11 deletion were 2/11 (18%) in fetuses with isolated RAA, 4/11 (36%) in fetuses with intracardiac anomaly. An extracardiac anomaly was observed in in 3/23 (13%) fetuses with isolated RAA, 6/29 (20%) in fetuses with intracardiac anomaly (total 10 fetuses, 19%). There were six pregnancy interruptions; two intrauterine deaths, 44 live births, two neonatal deaths and two patients were lost to follow-up. The RAA caused symptoms of vascular ring in one patient in the postnatal period.
Conclusion: The RAA has become more noticeable during fetal scans in recent years in spite of almost 50% of these being isolated cardiac lesions. Chromosomal and extracardiac anomalies are lower in isolated RAA but not negligible. The diagnosis of a right aortic arch can be made easily by non-paediatric/fetal echocardiographer whether it occurs as an isolated lesion or in association with other cardiac malformations. If the three vessels-trachea view is used as a routine screening method, the chances of picking up the right aortic arch in the fetus will be higher.

MP4-12

Twin-twin transfusion syndrome treated with fetoscopic laser coagulation: short-term cardiac function after birth

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Introduction: There are only a few reports on the effect of twin-twin transfusion syndrome (TTTS) on cardiac function in the infant. Since long-term cardiac outcome is grossly normal in TTTS survivors, but short-term studies are still lacking, we aimed to assess cardiac function in the early neonatal period to investigate whether short-term cardiac outcomes are also favourable.
Methods: Prospective echocardiographic follow-up of 93 TTTS pregnancies treated with laser surgery. Echocardiography was performed at the first day of life and at the age of 1 month (corrected for prematurity). Comparisons were made between donors and recipients and between TTTS-twins and a control-group of 9 uncomplicated monochorionic twin-pairs.
Results: 48 TTTS twin-pairs had both scans, 31 twin-pairs and 9 single survivors had one scan. At the first scan, donors had lower velocities across the aortic valve (maximum, mean and velocity time interval) as compared to recipients (maximum peak velocity: donor 0.65 ± 0.17 m/s vs recipient 0.74 ± 0.20 m/s, p=0.009), but not when compared to controls (p=0.96). As compared to

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

controls, both donors and recipients had lower tissue doppler imaging (TDI) derived indices of the left ventricle (S'-wave controls 0.049 ± 0.008 vs recipient 0.033 ± 0.013, p=0.00, vs donors 0.032 ± 0.011, p=0.00; A'-wave controls 0.053 ± 0.014 vs recipients 0.040 ± 0.0016, p=0.00, vs donors 0.043 ± 0.017, p=0.02). In donors the septal S'-wave was also decreased as compared to controls (0.035 ± 0.020 vs 0.039 ± 0.005, p=0.016). No significant differences in ventricular inflow or strain (rate) of the LV free wall were found. At the age of 1 month no (clinically) significant differences were found between donors and recipients or between TTTS twins and controls. Excluding infants with pulmonary stenosis (6 recipients and 1 donor) did not change the results.
Conclusions: TTTS survivors, both recipients and donors, show signs of left ventricular dysfunction at birth. In addition, lower velocities across the aortic artery in donor twins at the first day after birth could indicate poorer cardiac output in these infants. Cardiac function normalized as early as the age of 1 month in the majority of TTTS survivors.

MP5-1
Experimental Studies of Novel Transcatheter Flow Reducer in Pulmonary and Systemic Circulation

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Introduction: Flow reduction is sometimes needed to reduce pressure and prevent downstream vessel damage or to promote circulatory remodelling. We have recently reported preliminary data regarding the efficacy and retrievability of a new transcatheter flow restrictor (TFR; Occlutech GMBH, Germany) implanted in the systemic vein in a swine model. We report herein additional results obtained in the same animal model in which a TFR with refined shape was implanted in both systemic and pulmonary circulation.
Method: A refined prototype flow reducer (Occlutech GMBH, Germany) made of nitinol mesh with polyurethane covering was implanted percutaneously in the proximal left pulmonary artery (n=1) and in the intrahepatic segment of the IVC (n=2) in 3 domestic swine (weight 40-60 kg). The shape of the device was selected based on our computational fluid dynamics (CFD) study in order to optimize flow pattern and minimize risk of thrombosis. Following implantation, all animals were placed on warfarin as thromboembolic prophylaxis.
Results: The implantation procedure was uneventful with an initial pressure gradient of 4-5 mm Hg across the IVC device and 20 mm Hg across the LPA device, with no adverse changes in the systemic arterial pressure and oxygen saturation. All animals were euthanized 2 to 3 weeks after TFR implantation due to weakness and swelling of the left front limb which onset in all animals 2 weeks after TFR implantation without signs of increased venous congestion or heart failure. In one animal with device implanted in the IVC, catheterization prior to euthanasia showed good TFR patency with no pressure gradient across the device and moderate venous collateralization. In this animal, the device was easily retrieved using snare technique. The post-mortem examination of the animal with TFR implanted in the LPA showed fully patent device with moderate endothelization but no signs of neo-intima in the TFR's fenestration.
Conclusion: Transcatheter flow reduction in the systemic and pulmonary circulation using this device appears to be feasible with a smooth operator learning curve and possibility to retrieve the device weeks after implantation.

MP5-2
3D-printed endothelium-mimicking blood-contacting surfaces with anti-thrombogenic and anti-bacterial properties

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Background: Custom made, cardiovascular grafts are of advantage in the treatment of children with congenital cardiovascular defects in which the anatomy varies. 3D-printing is an inexpensive, accurate, and simple method which can help surgeons to design and print the grafts according to radiographical images. Thrombogenicity and bacterial infectiveness are the most common complications for foreign blood contacting surfaces associated with functional failure of the grafts.
Method: A 3d-printed model of blood vessel was designed and printed. A novel coating was designed to release NO in a controlled manner and in the physiological range of NO release comparable to that from the endothelium (0.5 - 4.0 × 10−10 mol.cm−2.min−1) to cover the 3d-printed graft. This coating was prepared by blending of S-nitroso-N-acetyl-D-penicillamine (SNAP) in a polymeric substrate created from polycaprolactone (PCL) and polyethylene glycol (PEG).
Results: Coating the 3D-printed grafts with PEG-PCL-SNAP resulted in quantitative anti-bacterial features against both Gram-positive and Gram-negative bacteria and in NO-mediated inhibition of platelet aggregation in the range of 14 days. Anti-bacterial and anti-thrombogenic properties in plasma are expected to be as effective as in PBS since NO release in plasma was not significantly different from that in PBS.
Conclusion: The application of NO-releasing form 3d-printed vascular grafts is promising for the engineering of vascular grafts showing bactericidal and anti-thrombogenic properties.

MP5-3
A chronic preload reduction animal (ovine) model: acute effects of reloading

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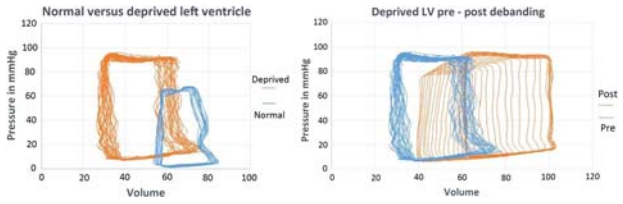
Introduction: Many cardiac conditions cause chronic volume deprivation of the systemic ventricle (e.g. Fontan, mitral stenosis, PAH, large ASD). We created a chronic volume deprived ventricle in an animal model (ovine) to study the effects of chronic volume deprivation and acute reloading.
Methods: in lambs a tight PFTE strip was placed around the inferior and superior caval vein through thoracotomy (n=14). Ten months later the PTFE bands were percutaneously dilated. Cardiac MRI was performed prior and within 48 h after debanding, hemodynamic data and PV loops (CD Leycom) were recorded prior and immediately (30 min) after debanding. Histology was done. Data was compared to age and weight matched healthy controls (n=9).
Results: 1/ Survival: 2 animals died after banding (ascites), 2 after debanding (rupture IVC).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



2/ Acute hemodynamic effects (PV loop): baseline EDP is elevated 9.0±3.3 mmHg compared to normals 1.0±3.4 mmHg (P <0.05). EDP rises after debanding to 12.4± 4.0 mmHg (P <0.05). The left ventricular CO is 3.3±0.7 pre and 3.0±0.6 L/min post debanding, compared to 3.0±1.2 L/min in normal animals. The EDV is 70.2+-8.7 pre and increases to 81.9+-13.5 ml post debanding (p <0.05), compared to 85±7.2 ml for normal animals. The ESV rises from 33.2±5.4 to 44.5±11.3 ml after debanding (55.6±18.4 ml in normal). Heartrate rises from 88±9 to 101±15 BPM compared to 94+-7 BPM in controls.
3/ Effect after 48 h (MRI): EDV on MRI is 70.5±7.7 ml before and 64.2±10.5 ml 48 h after debanding (P 0.79) ESV is 33.1±5.0 before and 34.1±11.3 ml 48 h later (P 0.14) EDV in healthy controls 76.1±14.1 ml and ESV 41.2±7.7 ml. 62.2±10.5 ml.
4/Histology: mean mass RV 29.2±4.4 g, LV 93.1±16.6 g was not significantly different from healthy controls; mean number of transected myocytes per 0.5 mm was RV 16.2±2.2 and LV 16.4 ±1.9, no signs of fibrosis.
Conclusion: In a chronic volume deprived ventricle the end diastolic pressure is elevated without marked histologic changes; EDP acutely rises when restoring the preload. Better understanding of this phenomenon may help avoiding/treating decreased ventricular compliance.

MP5-4
Multidisciplinary approach to Loeys-Dietz Syndrome in children
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Objective: The aim of this study is to describe the clinical manifestations of pediatric patients with Loeys-Dietz Syndrome followed-up at our center.
Methods: Epidemiology, genetics, clinical findings and follow-up were evaluated. All pediatric patients with Loeys-Dietz syndrome were included.
Results: A total of 14 patients were evaluated, with a median age at diagnosis of 10 years (range 0 to 20 years), 7 boys and 7 girls. Genetics: 4 (+1) TGFBR 1, 8 TGFBR2 and 1 TGFB2. Positive family history on 9/14 cases (64%). All patients presented aortic root dilatation except 2, median z-score of the aortic root +3.87 (range 0.53-7.11), and 9 also had aneurysms or dilatations at other levels. All the affected patients were treated with ARA-II and/or Beta-blockers. Aortic root replacement was needed on 6 (42%): 5 David's technique and 1 Bentall technique. Age range at surgery was 9 to 17 years old, and the range of Valsalva sinuses diameter was 38.5 to 47mm. One patient required pacemaker implantation immediately after David's surgery.
Half of the patients had pectus excavatum/carinatum, almost half had scoliosis and 5 had hindfoot deformity. Most common neurological involvement was tortuosity of the cerebral vasculature (7/14), and one patient suffered frontal hypoxic-ischemic events. Bronchiectasis were found on 2 patients, 2 obstructive sleep apnea and one spontaneous pneumothorax. Ophthalmologic involvement with strabismus or optometric disorders was described on 5 patients, and 1 retinal detachment. Other manifestations were one failure to thrive, one gastro-esophageal reflux, one food allergy and one asthma. Almost half of the patients had inguinal/umbilical hernias.
Median follow up of 4 years (range 2-9). Three patients died (21,4%). Two due to aortic dissection and one to cerebral hemorrhage at 12, 19 and 20 years of age.
Conclusions: Loeys Dietz Syndrome is a complex multisystemic disease. The prognosis is determined by the cardiovascular involvement, even in early ages. A multidisciplinary approach and a coordinated transition to adult units is mandatory.

MP5-5
The MFS-HARM score: prediction of cardiomyopathic features of Marfan syndrome with the MarFan-Syndrom-Heart-At-Risk-Model
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Background: Marfan syndrome (MFS) is a connective tissue disease frequently involving the cardiovascular system. Even though secondary cardiac manifestation in the form of valvulopathy is common, the question of primary cardiomyopathy still remains in debate. The purpose of this study is to understand the factors that predict cardiomyopathic features in pediatric MFS-patients.
Methods: Using data from 40 randomly selected MFS-patients from our pediatric Marfan database, echocardiography, and chart review, we developed a score predicting congestive heart failure (CHF) in patients with MFS. CHF was defined as either a cardiac index (CI) below the age adjusted lower limit of normal (LLNaa), a reduced ejection fraction (EF) below 50%, or the presence of clinical symptoms of CHF (NYHA >1). Using multivariable logistic regression analysis, predictors of CHF were determined and a score was postulated.
Results: In the 40 patients with secured MFS (age, mean±SD, 15.5±3.62 years) the CHF-incidence was 50%. Only 17.5% experienced symptoms of CHF (NYHA >1). The CI and the EF was on average lower in the CHF-cohort (mean±SD, CI 2.15±0.77 L/min/m2 vs. 3.03±1.29 L/min/m2, p=0.044; EF 60.62±12.81% vs. 67.72±11.76%, p=0.109). The presence of aortic valve regurgitation (22.5%) did not differ between the groups (p=0.268). Predictors of CHF were myopia >3 dpt, female sex, striae distensae, height <90th percentile, age <17 years, heart rate <LLNaa, and main pulmonary artery to sinus valsalva ratio > 0.7. Each predictor was assigned a point value of 1. The 7-point MarFan-Syndrom-Heart-At-Risk-Model (MFS-HARM) yielded a calculated area under the curve of 0.93. Each point increment was associated with a 8.27-fold increase in the odds for CHF (95% confidence interval, 2.44–28.00, p=0.001). An MFS-HARM score ≥3 predicts CHF with a 90% sensitivity and an 85% specificity.
Conclusion: In a randomly selected cohort of pediatric MFS-patients, 50% experience signs of CHF. The MFS-HARM, a composite score derived from patient characteristics tracked on follow-ups in a Marfan Center, predicts those at increased risk

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

for CHF. After validation, risk prediction along a continuum of clinical and imaging parameters may open opportunities to focus clinical resources on the prevention of the of cardiac function deterioration in adolescents with MFS.

MP5-6

First paediatric cohort for the evaluation of inflammation in endomyocardial biopsies

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Objectives: Endomyocardial biopsy (EMB) remains the gold standard for the diagnosis of myocarditis in children and adults. The existing WHO/ISFC criteria for lymphocytic cell infiltrates by Richardson et al. are based on myocardium of adults. The aim of this study was to analyse histopathological signs of inflammative myocardial disorder in a paediatric cohort.
Methods: The study prospectively enrolled patients <18 years with EMB, collected during a planned open heart surgery with routine resection of endomyocardial tissue from ventricular site. All patients had no history of infection or myocardial inflammation. The myocardium was formalin fixed and thereafter paraffin-embedded. For histopathological and immunohistological analyses 5-mm-thick tissue sections were stained with hematoxylin and eosin, Masson's trichrome, and Giemsa and examined by light microscopy. For immunohistological staining, monoclonal antibodies for the detection of T cells (CD3), B cells (CD20), macrophages (CD68) and major histocompatibility complex (MHC) II (HLA-DR) were used.
Results: Sixty-five endomyocardial samples from 65 patients were included. The myocardium derived from: 93.8% (n=61) right ventricular outflow tract, 4.6% (n=3) from the left ventricle and in 1.6% (n=1) from the right ventricle. The median patient age (interquartile range) at time of sampling was 0.6 (0.3-1.0) years, 66.2% male. A median of 2.5/mm2 (1.0-4.0) CD3+ T cells, 0.5/mm2 (0.0-0.5) CD20+ B cells and 4.0/mm 2 (2.0-4.0) CD68+ macrophages were detected. The MHC II grade was 0/mm2 in 16.9% (n=11), 0-1/mm2 in 53.8% (n=35) and 1/mm2 in 27.7% (n=18). All of these samples were below the current cut-off for myocarditis according to the WHO/ISFC criteria.
Conclusion: This is the first prospective study with analysing paediatric ventricular samples for the evidence of inflammative myocardial disorder. The degree of lymphatic cell in children without myocardial inflammation lies far below the existing thresholds in adults. Therefore detection of an increased number of lymphatic cells in EMB might already define a pathological inflammation in patients with suspected myocarditis.

MP5-7

Pulmonary hypertension after hematopoietic stem cell transplantation in children

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Background and objectives: Pulmonary hypertension (PH) is a rare but important cause of mortality after hematopoietic stem cell transplantation (HSCT) in children. We report here a series of children who developed PH after HSCT.
Population and methods: Over a period of 7 years, 367 children underwent HSCT (age range 0.5-252 months - median 20.3 months). After HSCT, cardiac echo scans, motivated by respiratory and/or hemodynamic symptoms, identified 31 patients with elevated tricuspid regurgitation velocity (> 2.8 m/s). Indications for HSCT were: 10 lymphohistocytosis, 11 SCID, 3 osteopetrosis, 1 Griscelli syndrome, 1 CD40 Ligand deficiency, 1 chronic granulomatose disease, 3 neuroblastoma, and 1 medulloblastoma.
Results: Seventeen patients had PH confirmed at right heart catheterization (RHC) (mean PAP=40.1±10 mmHg (range 28-62mmHg), PVRi=17.3±11.1 WU.m2 (range 8-42)). Vasoreactivity testing identified 13 responders according to Sitbon criteria. Five patients who were in too poor condition did not have RHC but were treated for PH according to echo signs. Nine patients did not have PH at RHC. Initial therapy was: calcium channel blockers in 6, oral monotherapy with PDE5i or ERA in 7, oral combo with ERA+PDE5i in 4, and up-front triple therapy with ERA+PDE5i+treprostinil in 5. Sequential add-on therapy was done in all 7 patients receiving mono- or oral combo. All patients receiving CCB needed add-on or switch for ERA and/or PDE5i. Seven/22 (32%) patients died: 6 of severe and progressive PH despite aggressive PH treatment, and one of infection after normalization of pulmonary pressure. Fifteen/22 PH patients are alive after a mean follow-up of 6.5±2.3 years. All survivors could be weaned of PH treatment after a mean follow-up of 6.2±3.1 months. The delay between clinical symptoms and initiation of PH therapy was significantly longer in patients who subsequently died (33.5±23 days-median 30) than in survivors (7±3 days) ($p<0.001$).
Conclusion: PH after HSCT is often misdiagnosed in the context of multiple co-morbidities. Interpretation of vasoreactivity testing is questionable and CCB non efficacious. Aggressive up-front combination therapy allowed frequent normalization of pulmonary pressure and improved survival. Systematic screening for PH after pediatric HSCT can be suggested to allow early detection and treatment.

MP5-8

Outcomes of peripheral pulmonary artery branch stenoses

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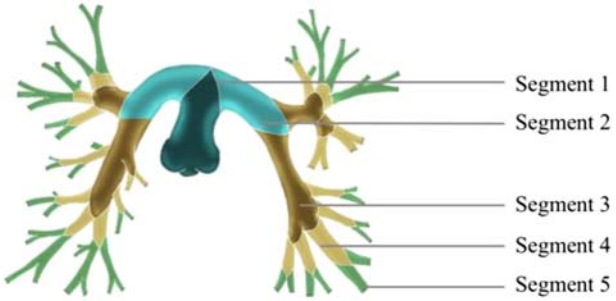
Background: Peripheral pulmonary artery branch stenosis (PPAS) is defined by the narrowing of pulmonary arteries after the hilum. Outcomes are scarcely known and therapeutic alternatives are limited.
Objectives: We sought to describe the different types of PPAS, and their respective outcomes.
Methods: Over a period of 10 years, all patients with PPAS were included into the study. Clinical, ultrasound and hemodynamic data were collected at the time of diagnosis. Only patients with biventricular repair of CHD were included. Patients were classified

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



into 3 groups (see figure): group 1 with diffuse hypoplasia from segment 1 to segment 4 or 5; group 2 with PPAS of segment 3 branches with or without aneurysms; and group 3 PPAS of segment 4 and 5.

Results: 45 patients were included: 11 group 1, 31 group 2, 3 group 3. One third had associated congenital heart disease. At time of diagnosis, 50% were under 5 years, and 80% were asymptomatic. Mean right ventricle/left ventricle (RV/LV) pressure was 0.99 and was not different between groups. 7 patients had percutaneous angioplasty and none was treated surgically. Median follow-up was 6.3 years. No changes in RV/LV pressure ratio was observed during follow-up. Survival at 10 and 20 years was 95.2% and 66.7% with no difference between groups. The presence of symptoms and RV pressure > 110 mmHg at diagnosis were significantly associated with right heart failure, hemoptysis, syncope and pulmonary branch thrombosis (p=0.003; p=0.04 respectively). Right heart failure and dyspnea on exertion at diagnosis were associated with death (p=0.01 for both).

Conclusion: PPAS is a rare and severe disease since one third of patients die in childhood. The anatomy of pulmonary branches did not predict outcome. The presence of symptoms and suprasystemic RV pressure are predictors of outcome.

MP5-9

Clinical presentation and survival of childhood hypertrophic cardiomyopathy associated with Friedreich ataxia: a national cohort study

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Background: The clinical spectrum of disease and survival in childhood hypertrophic cardiomyopathy (HCM) associated with Friedreich ataxia (FA) is poorly described. This study describes the clinical characteristics and outcomes of patients with FA associated HCM over four decades in a well-characterised United Kingdom (UK) cohort.

Methods: Demographic and clinical data for all children diagnosed with HCM secondary to FA between 1980 and 2017 were retrospectively collected.

Results: 74 patients with FA (39 male [53%]) met diagnostic criteria for HCM at a mean age 10.6 years (+/3, range 4.6-17.1). In 28 patients (38%) the diagnosis of HCM preceded that of FA. At the time of diagnosis: 24 patients (32%) were symptomatic (chest pain n=10; palpitations n=6; pre-syncope/syncope n=7; NYHA >2 n=12); 62 (84%) had concentric left ventricular hypertrophy with a mean LV maximal wall thickness 12mm (+/- 2.6, range 8-19mm); 1 (1.4%) had obstructive disease (LV outflow tract gradient >30mmHg); and 4 (5.4%) patients had impaired systolic function (EF < 50%). Over a mean follow up of 5.2 years (+/-4.7, range 0.5-28.7), 8 patients (12%) had documented supraventricular arrhythmias (Atrial flutter n=3, Atrial ectopic tachycardia n=1, Atrial fibrillation n=4, re-entry tachycardia n=1), 3 (4%) had ventricular arrhythmias (non-sustained ventricular tachycardia n=3) and 1 (1.4%) had conduction disease. Freedom from supraventricular arrhythmias was 98.6 (95% CI 90.7-99.8) at 1 year and 74.7% (95% CI 49.4-88.7) at 10 years. 39 patients (53%) were started on medications for cardiac symptoms (B-blockers n=16, Calcium channel blocker n=7, heart-failure medications n=5, Anti-arrhythmics n= 4, other n= 7), 1 underwent a cardiac transplantation (aged 4 yrs.), 1 suffered a transient ischaemic attack (aged 13 yrs.) and 4 patients (5.4%) died (congestive cardiac failure n=1, atrial arrhythmia-related n=2, non-cardiac n=2). There were no sudden cardiac deaths. Overall mortality rate was 1.35 (95% CI 0.61-3.01)/100 patient years follow up.

Conclusions: This national study of FA-associated childhood HCM is the largest reported and describes a symptomatic cohort of patients with a high prevalence of childhood atrial arrhythmias and early progression to end stage disease. Overall mortality is similar to that reported in non-syndromic childhood HCM but no patients died suddenly.

MP5-10

Electrocardiogram findings in Pediatric Patients with Myocarditis

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Introduction and Objectives: Acute myocarditis(AM) is an inflammatory illness caused by a myocardium infectious process and/or immune response. Electrocardiograms(ECG) abnormalities are common in patients with AM and some of them have been reported as predictors of poor clinical outcome in adults.

The objective of this study was to describe initial ECG abnormalities of pediatric patients with AM, and its relationship with clinical outcome.

Methods: We analyzed the initial ECG of all pediatric patients (0-16 years) with AM (diagnosed by endomyocardial biopsy (EMB) or cardiac magnetic resonance(CRM)) who were admitted in our hospital from April 2007 to December 2018. ECG values were compared with normal reference values for group age.

The association between ECG changes left ventricular ejection fraction(LVEF) < 35%, presentation of cardiogenic shock, need for ECMO, transplant and mortality, was analyzed.

Results: 41 patients with 42 myocarditis episodes were included. 68% males, with a median age of 25 months (7days to 16years). The diagnosed was made by EMB in 14 and by CRM in 28 patients.

ECG at admission was abnormal in all the patients. Abnormalities included: repolarization anomalies with T wave inversion (46%), widening of QRS (41%), wide QRS-T angle (greater than 100°)

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

(36%), reduced voltage (35%), pathological ST elevation (32.5%), prolonged QTc (10%), presence of pathological Q waves (7.5%). ECG findings associated with cardiogenic shock were the presence of pathological Q waves (100% vs 37%: p=0.03) and wide QRS (62% vs 30% p=0.047). The rest of the abnormalities did not have any statistically significant association. Patients with pathological ST elevation without Q waves had a more benign clinical course with a higher LVEF and a complete recovery in 100% of the cases.
Conclusion: All pediatric patients with AM had ECG abnormalities at admission, therefore, the finding of a normal ECG nearly rules out myocarditis as a diagnosis.
The presence of pathological Q waves and widening of QRS was associated with a more severe clinical presentation.
Wide QRS-T angle, described in adults as poor prognosis predictor, did not correlate with cardiogenic shock, lower LVEF, higher mortality or transplantation.
Patients with pathological ST elevation without Q waves had an excellent prognosis.

MP5-11
Afterload augmentation is driven by the preload reduction rather than venous congestion in the chronic Fontan circulation

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Background: Based on the Pressure-Volume framework in the Fontan circulation, enhanced cardiac afterload compromises stroke volume, which exerts beta-adrenergic and RAAS cascades, ultimately resulting in cardiovascular remodeling. Since venous congestion directly accelerates end-organ dysfunction, early establishment of operable circulation with minimal central venous pressure (CVP) by balancing ventricular loading conditions (preload/afterload) would be the key to avoid "Fontan failure". We hypothesized that venous congestion augments cardiac afterload in the Fontan patients.
Method: Transient inferior vena-caval occlusion (IVCO) was applied to induced venous congestion and preload reduction in 110 patients (23 Fontan and 87 non-Fontan), and augmentation of arterial elastance (Ea) was measured under construction of ventricular pressure-volume relationship. In addition to load independent measures of ventricular function, mean circulatory filling pressure (mcfP) was estimated and neurohormonal activations were evaluated.
Result: As compared with non-Fontan patients, lower cardiac output in the Fontan patient was attributed by the augmented afterload with similar contractile function. While induced CVP augmentation at proximal to the IVCO was markedly higher in the Fontan patients as compared to non-Fontan patients (p=0.0017), increase of Ea during IVCO was markedly suppressed in the Fontan patients (p=0.0090). Interestingly, while Ea augmentation in the non-Fontan patients was affected both by venous congestion (p=0.0091) and preload reduction (p=0.0011), that in the Fontan patients was solely but markedly augmented by the preload reduction (p<0.0001, ANCOVA: p=0.049 vs non-Fontan). To further delineate interaction between congestion and afterload, association with mcfP and systemic vascular resistance (SVR) was analyzed. Ea augmentation was negatively correlated with mcfP (p=0.025) as well as systemic vascular resistance (p=0.047), suggesting importance of decongestion for the preservation of Ea augmentation. Consistent with this, Ea augmentation was negatively correlated with natriuretic peptides (ANP/BNP), while serum level of aldosterone was independent of it.
Conclusion: In the chronic Fontan circulation, insufficient preload, rather than additional venous congestion, predominantly increased afterload, implied vital role of preload preservation. In contrast, Ea agumentation against venous congestion was preserved in the non-Fontan patients implied need for optimizing mcfP by volume and venous manipulations.

MP5-12
Inspiratory ventilatory training in patients with repaired tetralogy of Fallot

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Objectives: Tetralogy of Fallot (TOF), a right heart obstructive congenital heart disease (CHD) affects the pulmonary system. The undersupply in the lungs may lead to lower exercise capacity and impaired lung volumes. Training of deep inhalation may train the lung volumes, improve pulmonary blood flow and lead to a better exercise capacity and lung function. This study examines the effects of a volume-oriented inspiratory ventilatory training in patients with repaired TOF on exercise capacity and lung volumes.
Methods: From February 2017 to November 2018 fifty-four patients (age at inclusion: 14.7 ± 4.8, 39% female) completed a ventilatory training. All of them had a TOF or a similar CHD. They underwent a spirometry (forced vital capacity, FVC; forced expiratory volume within the first second, FEV1) and a cardiopulmonary exercise test (CPET: peak oxygen uptake, VO2peak). Data were compared to references and expressed as %predicted (mean ± SD). Statistical analyses were done via Student's t-Test for dependent samples. Patients were re-examined six month after their training.
Results: While VO2peak did not significantly increase after the training, lung function improved (FVC: 83.6 ± 2.2 vs. 86.1 ± 2.4%predicted, p=0.004; FEV1: 82.5 ± 2.2 vs. 82.5 ± 84.8%predicted, p=0.017). FEV1 to FVC ratio did not change significantly.
Conclusions: This study shows that patients with repaired TOF benefit from a volume-oriented inspiratory breathing training with regard to their spirometry results. Exercise capacity did not show any improvement.

P-1
Preliminary experience with a novel High Density Grid Mapping Catheter for cardiac mapping in patients with and without congenital heart disease

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Introduction: Cardiac mapping is time-consuming and frequently incomplete. The idea of high-density mapping is to obtain a more comprehensive understanding of the arrhythmia mechanism and substrate to facilitate the ablation procedure and to enhance ablation success.
Methods: 3D non-fluoroscopic electroanatomic cardiac mapping (Ensite NavX Precision, Abbott) was performed utilizing a novel high-density mapping catheter (Advisor HD Grid Mapping Catheter, Abbott) followed by conventional point-by-point

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Table 1

Pt-No	CHD	Substrate
1	DILV, Fontan with dilated lateral tunnel	Multiple IARTs
2	TA and PA, Fontan with dilated tunnel	Multiple FAT and IARTs
3	DILV, Fontan	IART
4	TA and PA, Fontan with lateral tunnel	FAT
5	ASD, closure with patch	multiple IARTs
6	DORV, CoA, VSD closure with patch, subclavian flap, RVOT patch	Monomorphic RVOT-PVC, incessant RVOT-VT
7	PA- VSD, RV – PA conduit	RVOT-VT, polymorphic RVOT-PVC
8	D-TGA, Senning, baffle leak	Multiple IARTs
9	None	Idiopathic RVOT-PVC/ns VT
10	None	Left lateral AP
11	None	Idiopathic RVOT-PVC
12	None	Left posterolateral AP

mapping in areas of interest. A paddle-like formed distal tip of the 8-F catheter is created by four soft splines. 16 electrodes are distributed over the four soft splines (1 mm electrode length separated by 3-3-3 ring spacing). The equidistant spacing allows a bi-pole recording along and across the splines.
Results: Our single center study comprised 12 patients (table 1; age 25 +/- 11 years, range 7 to 46 years) undergoing ablation of supraventricular tachycardias, premature ventricular contractions (PVC) or ventricular tachycardias (VT). Primary ablation success was achieved in all patients (endpoints defined as: non-inducibility n = 6, bidirectional conduction block n = 2, absence of AP-conduction n= 2, absence/non-inducibility of PVC n = 2).
Conclusion: The novel HD mapping catheter appears to facilitate cardiac mapping (activation and substrate) and catheter ablation, especially in patients with congenital heart disease. Further studies are needed to evaluate whether this approach improves primary and long-term catheter ablation success.

P-2
Fever In Children At-Risk For The Brugada Syndrome

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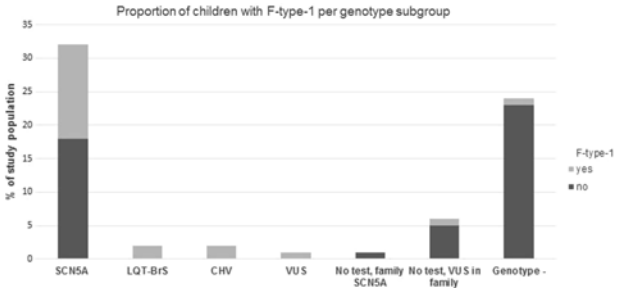
Introduction: (or Basis or Objectives): Brugada syndrome (BrS) is a rare inherited arrhythmia syndrome. Of the genes associated with BrS, variants in the SCN5A gene are most frequently described. Usually however, no associated genetic variant is identified. BrS patients, especially children, are at risk for arrhythmic events during fever. Fever is known to unmask the Brugada-type-1 pattern. Children with a positive family history for BrS, genotype-positive, and phenotype-positive children are considered at-risk for Brugada-type-1 pattern and cardiac events (CE). They are thus advised to record an ECG during fever, which is a stressful experience.
To evaluate the current policy, we aimed to identify risk factors for Brugada-type-1 pattern during fever (F-type-1) and CE by evaluating demographics and genetic background in an at-risk paediatric population.
Methods: Children with a positive family history for BrS, genotype-positive, and phenotype-positive children with ≥1 available fever-ECG from three tertiary medical centres were included and divided into two groups depending on the occurrence of F-type-1 during follow-up. Demographics and ECGs during fever were retrospectively retrieved and analysed. Chi square test and applicable post-hoc analyses with Bonferroni correction were used.
Results: The study population consisted of 68 children, 21 were allocated to the F-type-1 positive group and 47 to the F-type-1 negative group. The groups differed significantly in CE; in the F-type-1 positive group 7 (33,3%) children had CE during follow-up in comparison to 0 in the F-type-1 negative group (p=0,000). Five of them had symptoms during fever. The two groups also differed significantly in genetic background (p=0,002, See Figure 1). In the F-type-1 positive group 14 (43,8%) were SCN5A variant carriers in comparison to 1 (4,2%) in the genotypic negative children (p=0,001). When children diagnosed with BrS at first presentation after CE (n=3) or type-1-ECG at baseline (n=1) were excluded, this result remained unchanged.
Conclusions: Our results suggest that carrying a SCN5A variant is a risk factor for F-type-1 in children. F-type-1 is also associated with symptoms in this population. Children carrying SCN5A variants associated with BrS should be closely watched during febrile episodes and an ECG should be recorded.



P-3
ECG as a predictor for coronary artery involvement in Kawasaki disease

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Introduction: Kawasaki disease (KD) is a pediatric acute and self-limited multisystemic vasculitis of unknown etiology that mainly affects small and medium-sized arteries, constituting the most common cause of coronary heart disease. Some studies related ECG findings with coronary disease in this population. We aim to analyze whether ECG could be used to predict coronary artery involvement (CAI).
Methods: In this retrospective study including patients diagnosed with KD (AHA criteria) with available ECG data obtained during the first month of KD diagnosis. Patients with underlying disease that would have altered the baseline ECG were excluded.
12-lead-derived ECG analysis was performed using a hand calibrator, 11 ECG variables and T wave pattern morphologies. The Chi-square test and/or the Fisher exact test were used for the categorical variables. The Mann-Whitney U test was used for comparisons of quantitative variables. Values of $p<0.05$ were considered statistically significant. Values are expressed as median[IQR].
Results: 50 patients met KD criteria, 19 were excluded and 31 were included for analysis. Age at diagnosis was 36 months. Twelve (38%) had CAI. Patients with CAI had similar ECG results compared to the non-CAI patients, QTc interval (413 [52] vs 420[58]), Tp-e/ QTc (0.18[0.05] vs 0.17[0.06]), T amplitude (0.32[0.21] vs 0.3[0.18]) QT interval (280[30]) vs 280[80]) Tp-e (80[10] vs 80 [20]), Tp-e/QT (0.28[0.05] vs 0.25[0.09]) and T wave duration (160[44] vs 140[40]) were longer but this differences did not reach significance. CAI patients had more IVIG resistance compared to the non-CAI patients (6/12[50%] vs 5/19 [26%]).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusions: ECG repolarization pattern showed changes during acute and subacute phase of KD. However, no statistically significant correlation was observed between these changes and the coronary artery involvement or IVIG resistance. Future studies encompassing larger series are needed to evaluate whether ECG could be use to predict CAI in KD.

P-4

Radiofrecuency Catheter Ablation of Supraventricular Tachycardia in a Preterm Infant. Case Report

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Introduction: Sustained fetal tachycardia is an uncommon situation. It may cause fetal hydrops, preterm delivery, and higher perinatal morbidity and mortality. Accessory pathway is the most frequent cause of tachycardia in neonates and is usually well controlled with antiarrhythmics. The risks of radiofrecuency catheter ablation are still high in infants because of their low weight and the small size of their vessels, so it is reserved for arrhythmias without medical treatment response.
Case Report: Fetal tachycardia with heart rate 230 bpm detected in a routinary echocardiography in week 32. Hydrops was present. Trans-placental treatment with digoxin and flecainide was started, achieving a heart rate decrease (200 bpm) and partial resolution of hydrops. Spontaneous delivery occurred at 32+6 weeks. At birth, ECG showed regular tachycardia, 220 bpm, with narrow QRS and short RP. Echocardiography was normal except for a severe mitral regurgitation (also present prenatally). Sinus rhythm was accomplished with several doses of endovascular adenosine but only for a few seconds. Treatment with digoxin and propranolol was started with no results. Different drug combinations were used unsuccessfully: flecainide, digoxin + flecainide, propranolol + flecainide, amiodarone, and amiodarone + digoxin + propranolol. Patient continued hemodynamically stable although echocardiography showed mild heart dysfunction. At 19th day of life, with 2.5 kg of weight, electrophysiology study was performed under general anesthesia and through a right femoral vein approach. Patient presented sustained tachycardia with narrow QRS, right bundle branch block and negative P wave in aVL, II, III and aVF. Left atrium was accessed through the foramen ovale; a mitral ring cartography was performed with 6 French catheter. Ablation was performed with radiofrequency (25 watts) of a left accessory pathway (in the lower mitral ring area) with orthodromic AV conduction. There were no intra-procedure complications. Two years later the patient remains stable with sinus rhythm and normal heart function without any medication.
Conclusions: Radiofrequency catheter ablation may be a therapeutic option in preterm infants with incessant supraventricular tachycardia. It may be the technique of choice in refractory cases with hemodynamic compromise.

P-5

Catheter ablation for tachyarrhythmia in children. The impact of Electroanatomic Mapping Systems on Fluoroscopic Exposure Reduction

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Objective: Retrospective study aimed to describe the results of catheter ablation for SVT in children<17 years at a single institution during the period 2013–18 and to compare the safety and efficacy of procedures with and without electroanatomic mapping system guidance.
Patients and Procedures: 98 consecutive patients who underwent a total of 106 catheter ablation procedures were included (9 with cryoablation and 97 with radiofrequency energy). Median age was 11,1 years (range 0,1 to 16,9 years) and median weight was 45 kg (0,3 to 80 kg). 57 were males. Congenital heart disease was present in 6 patients. Arrhythmia substrates was Wolff–Parkinson–White syndrome in 42, concealed accessory pathway in 27, atrioventricular nodal re-entry tachycardia in 23, permanent junctional reciprocating tachycardia in 3, focal atrial tachycardia in 3. 63 procedures (group 1) were performed only under fluoroscopic guidance and 43 (group 2) using an electroanatomic mapping system (Carto or NavX).
Results: The acute success rate was 95% and 98% when including repeat procedures. Arrhythmia recurrence occurred in 5 patients (5%). 3 complications were observed: 1 pericardial effusion and 2 Wenckebach atrioventricular block that spontaneously regressed. The acute success did not differ between group 1 and 2 (p 0,629) and no difference was registered in procedure time (median 104 versus 100 minutes, p 0,7). Complication rate did not differ significantly between groups (p 0.56), although the incidence was higher in group 1 (3 cases versus 0 in group 2). Fluoroscopic exposure was significantly reduced in group 2 compared to group 1 (median 0,7 versus 11,2 minutes; p<0,001). Catheter ablation was completely performed without fluoroscopy in 17 patients.
Conclusion: Catheter ablation can be undertaken in children with a high success rate, few recurrences and complications. Non-fluoroscopic electroanatomic mapping can significantly reduce the radiological exposure. This is especially important in children as radiation can be potentially more harmful in younger individuals.

P-6

Videothoracoscopic Bilateral Sympathetic Denervation in Children with Long-QT Syndrome and Catecholaminergic Polymorphic Ventricular Tachycardia

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Objectives: Left cardiac sympathetic denervation (LCSD) is a good proven surgical option as adjunct therapy for patients with life-threatening ventricular arrhythmias, in reducing arrhythmic events and frequent ICD shocks. Performing bilateral CSD (BCSD) is reported to be more effective and with similar complication rates when compared to LCSD, but there is only a few data about pediatric cases in the literature. We aimed to present the initial outcomes of cases with BCSD performed due to long QT syndrome (Long-QTS) and Catecholaminergic polymorphic ventricular tachycardia (CPVT) in our center.
Patients and Methods: We retrospectively reviewed the electronic medical records of our pediatric cardiac arrhythmia center for all patiens with a diagnosis of Long-QTS (n=117; 22 with an

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

ICD) and CPVT (n=19; 15 with an ICD), and found a total of 11 cases with BCSD operation. The demographic features, operation data, medical treatment, ICD records and follow-up data for noticed. In all cases, bilateral T2-4 sympathetic ganglions and Kuntz fibers were cauterized videothoracoscopically.
Results: Among the 11 cases(5 female), 5 had long-QT syndrome and 6 CPVT. Mean age of the patients was 12.45 years(4-17). Seven cases had a 2-port and 4 cases a 3-port thoracoscopic BCSD operation. The mean operation duration was 45,63 minutes (38-65). Mean postoperative hospital stay was 1.63 days (1-3). Apart from one CPVT case with polymorphic VT during the operation (returned to sinus after defibrillated externally) and one case with pneumothorax (lasting two days), no additional complication or mortality was observed. Mean follow-up after procedure was 17.6 months. In the cases with an indication to reduce recurrent ICD shocks(n=9), mean preoperative shocks of 14.70/year fell to 1,09/year. In the other two patients with an indication of primary prevention (in one case who rejected ICD implantation, and in the other case after extraction of the ICD, due to infection) follow-up continued without any arrhythmic event.
Conclusion: Directly performing BCSD appears to be effective and safe in reducing life-threatening arrhythmias and ICD shocks in pediatric CPVT and Long-QTS. But we need further studies to compare it with LCSD for efficacy and safety.

P-7
Idiopathic ventricular tachycardia originating from the left/right coronary cusp commissure in two pediatric patient; A unique entity with specific electrophysiologic features

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Idiopathic ventricular tachycardias (IVT) originating from the left/right coronary cusp (L-RCC) commissure are rare. They have some unusal electrophysiologic features, and only a few reports from adult cases are found in the literature. We present two pediatric cases, ablated with radiofrequency(RF) energy.
Case-1: A 9-year old, 26 kg girl was referred to our hospital with symptomatic sustained VT attacks, and syncope. VT was stopped with electrical cardioversion during the last attack, because of hemodynamic instability under medical therapy with metoprolol, diltiazem and amiodaron infusion. ECG characteristics of VT were a tachycardia cycle lenght(TCL) of 330 ms, wide QRS complexes characterized by left bundle branch block (LBBB) morphology, positive in inferior leads and lead D-I, transition zones between precordial leads V3 and V4 and maximum deflection index (MDI) of 0.74. Transthoracic echocardiography showed no structural pathology and normal ventricular contraction. Interestingly, Holter-ECG monitorization didn't reveal any premature ventricular contraction (PVC). During the electrophysiologic study (EPS), she had no PVCs and sustained VT was induced with orciprenaline and dobutamine infusion. During activation mapping of VT in right and left ventricular outflow tracts (RVOT and LVOT), the earliest local endocardial activation sites (ELEAS) were found at posteroseptal region of RVOT with -32 ms and at the L-RCC commissure in LVOT with -62 ms(Figure1-A). The focus was ablated successfully with 5F RF-catheter using 25 watt energy(Figure1-B), and above the aortic valvar anulus plane, an unusual location for most aortic cusp VTs.

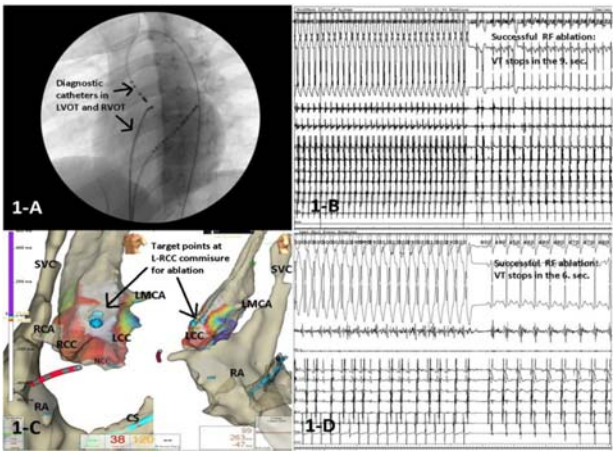

Figure-1.
A. *Flouroscopic image from the first case showing two diagnostic catheters in RVOT posteroseptal and LVOT L-RCC comissure regions with ELEAS values, very close to each other.*
B. *Intracardiac ECG records from the first case during successful RF ablation, showing a'burst' VT acceleration and abrupt cessation in the 9. second of the first RF energy application.*
C. *3-D electroanatomic image from the second case showing target lesions at L-RCC commissure with ELEAS for ablation, and their relation to LMCA.*
D. *Intracardiac ECG records from the second case during successful RF ablation.*

Case-2: A 16-year-old girl admitted to our department with recurrent palpitations and documented VT during her last attack, terminated with amiodarone infusion. ECG properties were similar to first patient with TCL:310ms and MDI:0.68. Again, holter monitorization revealed no PVCs. During activation mapping of VT, ELEAS were found at posteroseptal region of RVOT with -37 ms and at L-RCC commissure in LVOT with -65 ms (Figure1-c). It was ablated successfully with 7F RF-catheter, using 25 watt energy(Figure1-D).
In conclusion: IVTs originating from L-RCC commissure have specific features including absence of premature ventricular contractions, high MDI index suggesting epicardial origin, and ELEAS values for typical RVOT- VTs on EPS, causing to misinterpretation unless a LVOT-mapping is done.

P-8
Succesfull RF ablation for the treatment of electrical ICD storm in a child with early repolarization and idiopathic ventricular fibrillation

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Early repolarization(ER) may sometimes be associated with ventricular fibrillation(VF) and sudden cardiac death. A pattern of convex upward J wave, with ST-segment changes, especially in inferolateral leads on Electrocardiography(ECG) suggests "malignant" ER. We report a 14-year-old boy with ER associated idiopathic VF and electrical ICD storm controlled by intravenous isoproterenol and successfully treated by radiofrequecy(RF) ablation finally.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

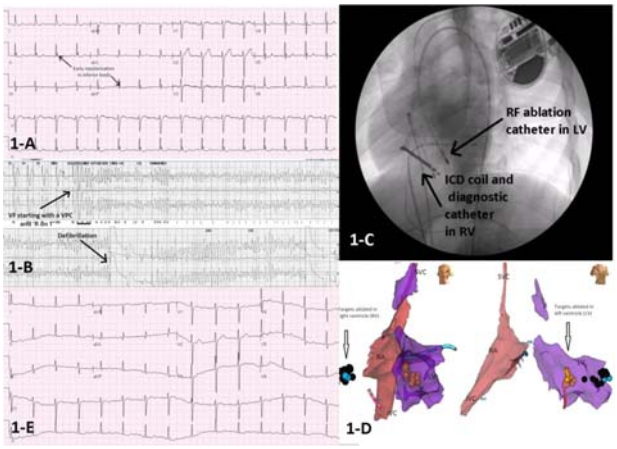

Figure-1.
A- Twelve lead surface ECG on admission with 'malignant' early repolarization signs in inferior leads.
B- *ECG record of ventricular fibrillation and ICD shock.*
C- *Flouroscopy image showing iirrigated RF ablation catheter in LV and diagnostic catheter in RV, beside the ICD coil.*
D- *3-D Map showing ablated Purkinje potentials in both ventricles; in RV shown as black and in LV as yellow.*
E- *12 lead Surface ECG with almost lost of ER sign.*

The patient was referred to our clinic with aborted sudden cardiac arrest, while resting at home. ECG yielded malignant type ER pattern and no significant structural defect was noticed on his echocardiograpgy(Figure1-A). Coronary angiography, cardiac magnetic resonance, epinephrine-ajmalin challange tests and electrophysiologic study(EPS) revealed no specific pathology. After that he was implanted a transvenous ICD(Medtronic-Evera, MRI-SVR Sure-scan) for secondary prevention and Metoprolol was initiated.
After 2 monts, he admitted to our ICU unit because of ICD storm with more then 20 shocks in a day(figure1-B). On İCU first day he further experienced a lot of appropriate ICD shocks, despite multiple antiarrhythmic medications(combination of beta bloker plus amiodaron, flecainide and mexiletine). Finally we strated intravenous isoproterenol infusion and VF storm was controled. Amiodarone plus oral disopyramide was initiated as maintenance therapy (because quinidin sulfate is not found routinly in our country). After an attempt to discontinue isoproterenol, the patient had further VF attacks and mutiple ICD shocks, so we decided for an urgent EPS and catheter ablation with ECMO backup ready. During the study we couldn't stimulate any premature ventricular contraction or VF, so we ablated local purkinje signals in both ventricles with an irrigated RF catheter (Figure1-C and-D). Especially after the left ventricular purkinje signal substrate ablation, ER almost disappeared on inferolateral leads(Figure1-E). After one week of successful ablation, the patient was discharged with oral amiodaron and disopramide therapy. He is still on follow-up without any VF attack and ICD shocks.
In conclusion; this case report represents the efficiency of the isoproterenol infusion for acute control of idiopathic VF associated with ER and electrical storm. Hovewer, if recurrent VF attacks and İCD shocks continue or recure after cessation of isoproterenol infusion, purkinje network ablation should be keeped in mind, even in children.

P-9

Catheter ablation of idiopathic fascicular ventricular tachycardia in children; Five years single center experience

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Objectives: Idiopatic fascicular ventricular tachycardia(FVT) is characterized by relatively narrow QRS tachycardia with right bundle branch block on ECG. It generally affects young-male population. It has a reentrant mechanism using Purkinje network as a one of its components. The data of the FVT ablation technics and results in childhood is still limited. The aim of this study is to present our experience in children with FVT ablation.
Patients and Methods: We reviewed retrospectively records of 945 transcatheter electrophysiological study(EPS)-ablation procedures done in our clinic between November 2013-November 2018, and found a total of 128 patients ablated for ventricular arrhythmias (PVC/ VT) ablation. Among them, 19 patients ablated with posterior fascicular VT diagnosis were included the study.Catheter ablations were performed with EnSite-3D mapping system guidance and limited fluoroscopic exposure.
Results: The mean age was 12.6±4.0(1.4-19.5) years, and the mean weight was 50.0±20.8(10.5-100) kilogram.Fifteen patients(78.9%) were male. Palpitations(89.4%) and fatigue(15.7%) were the most common symptoms. Interestingly, 6 patients(32%) were referred to our center with the diagnosis of supraventricular tachycardia with bundle branch block. False tendon was found in 8 patients (42.1%).According to ECG findings; posterior FVT 17(89.4%) and anterior FVT 2(10.6%) was diagnosed.
During the EPS, FVT was stimulated with basal stimulation in 7 patients, and medication(orciprenaline and/or dobutamine) was required in 12. Reentrant FVT was most frequently inducted with proximal coronary sinus catheter atrial stimulation(47.3%).Among patients with posterior FVT;typical-AVNRT(n=2) and upper-septal FVT(n=1) were also stimulated.Average VT cycle length was 322±66 ms(200-450) and VT was sustained during EPS in 15 patients(78.9%). Mapping and ablation was performed was performed during VT in 13 patients(68.9%), remaining 6 were ablated in sinus. Mean number of radiofrequency application was 8.2±3.9 (3-15).The mean fluoroscopy time was 4.5±1.9(2.1-8.5) minutes and the mean procedure time was 151.9±39.0 minutes (range, 102-240).There was no complication. Ablation was acutely successful in all (19/19, 100%) cases.The mean follow-up period after ablation was 19.8±16.1(2-48) months and VT recurred in one patient. Therefore the cumulative ablation success was 94.7% (18/19).
Conclusion: Radiofrequency catheter ablation is an effective and reliable method for the treatment of IFVT in childhood. Despite difficulties of VT induction during EPS, recurrence rate is relatively low.

P-10

Left coronary cusp cryoablation of the accelerated idioventricular rhythm in a child, triggering torsades de pointes and resulting in cardiac arrest

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Accelerated idioventricular rhythm (AIVR) is known as a benign arrhythmia and normally requires no specific treatment. But very rarely, it can also give rise to more severe arrhytmias. Here, we present a child with left coronary cusp-originating AIVR, degenerating in to Torsades de pointes (TDP) and resulting in cardiac arrest, which was ablated with a cryocatheter. To our knowledge this is the first pediatric case in the literature.

An 11 years-old boy, who has been followed due to asymptomatic AIVR in another center before, was referred to our department, because he had experienced an aborted cardiac arrest during sleep. He had been resuscitated for 5 minutes. 12 lead surface ECG showed frequent AIVR, with a left bundle branch block morphology, QRS axis in inferior leads and a QRS transition zone on precordial V3(Figure1-A). A 12 lead 24 hour Holter-ECG revealed incessant AIVR, consisting up to 90% of the whole record and two TDP attacks, triggered by AIVR–induced 'R on T' phenomenon, and resulting in syncope and cardiac arrest (figure 1-B). The exercise treadmill test was normal. Transthoracic echocardiography revealed no structural cardiac defect but mild left ventricular sistolic dysfunction with an EF of 45% and SF 23%. An electrophysiologic study (EPS) was conducted under general anesthesia. AIVR focus was mapped to left aortic coronary cusp (figure 1-C), and selective left coronary angiography revealed close neighborhood to coronary ostium, so a cryocatheter with an 8 mm tip was prefered for successful ablation of the AIVR focus (Figure1-D). The patient was discharged in three days without any PVCs or AIVR and with normal cardiac functions (EF 66% and SF 35%). After 9 months on follow-up, he is still asymptomatic, without any PVCs/AIVR,normal ECG (QTc:412 ms), and normal cardiac functions.

Although the clinical course of AIVR is known as benign, this case report shows that it may also degenerate to a life-threatening arrhythmia. EPS and catheter ablation is a good option in such cases for an ultimate cure. This case also showed that; cryoablation is a good and safe alternative to RF ablation in procedures close to the coronary arteries.

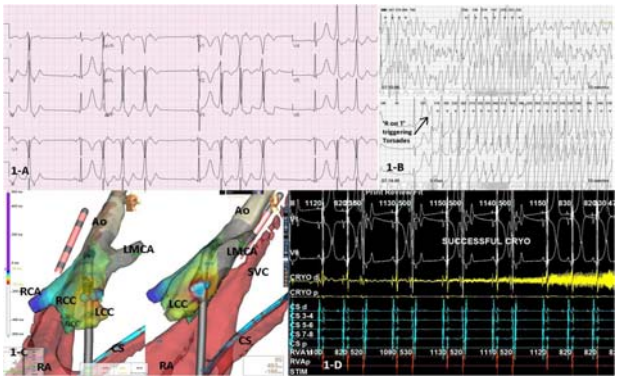

Figure 1.
A- Twelve lead ECG with frequent AIVR, B-'R on T' phenomenon triggering Torsades on Holter-ECG C- 3-D Map showing the focus in the left cocnary cusp, near the orifice of LMCA, ablated with Cryocatheter.C- İntarcardiac ECG showing successful cryoablation of the frequent AIVR.

P-11

Successful right anteroseptal manifest accessory pathway cryoablation in a six month infant with dyssynchrony-induced dilated cardiomyopathy

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Wolff-Parkinson-White (WPW) syndrome with right-sided septal or paraseptal accessory pathways (APs) may cause eccentric septal activation, resulting in dyssynchrony and left ventricular dysfunction. Catheter ablation of the AP is an effective treatment option, although with high complication rates in infants. We present a case of six month age infant, who was diagnosed with WPW syndrome with right-sided anteroseptal (parahisian) AP and dilated cardiomyopathy.

She was referred to our hospital with heart failure symptoms, weighing 8 kg, and having grade 1-2/6 heart murmur, mild tachypnea and hepatomegaly on physical examination. There were typical ventricular preexcitation signs like short P-R interval and wide QRS complexes with delta waves on 12-lead surface ECG, suggestive with an anteroseptal manifest AP (Figure 1A). Echocardiography showed rightward systolic bulging of the basilar septum and a dilated left ventricle with impaired systolic functions ((LVEDD:40 mm (Z-score:+ 4.4) and ejection fraction (LVEF) 34% by Simpson's method. Measurements for interventricular mechanical delay (IVMD) and septal-to-posterior wall motion delay (SPWMD) were 74 ms and 290 ms respectively, consistent

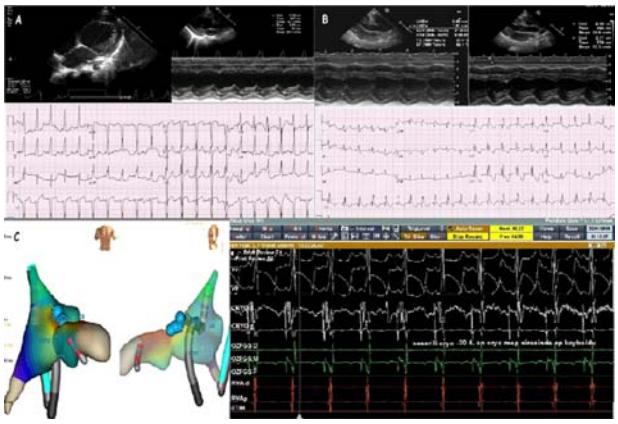

Figure 1.
A: Decreased LV systolic function with an EF of 34,6% (Simpson's). M-Mode ECHO showing marked dyssyncrony caused by the ventricular preexcitaion of the anteroseptal AP. 12 lead surface ECG on admission, showing WPW preexcitation.
***B:** M-mode echocardiograpgy showing improved left ventricular sistolic function, with an EF of %69. 12 lead surface ECG, showing no ventricular prexcitaion sign and incomplete RBBB after successful ablation of the AP.*
***C:** 3D anatomy of the right atrium and ventricule, with blue dots showing targets on the anteroseptal region, and diagnostic catheters in high rigt atrium (HRA), esophagus (OZFGS) and Wright ventricle (RVA), and the 6mm cryocatheter active, in the middle. İntracardiac and surface 12 lead electrograms during the successful cryoablation.*

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

with dyssynchrony (Figure 1A). Because of symptomatic dyssynchrony-induced dilated cardiomyopathy, electrophysiologic study (EPS) was performed.The patient was intubated and the EPS was performed under general anesthesia. Right and left fomaral veins were catheterized and also an esophageal catheter was inserted. 3-D mapping and floroscopy were used together during delta mapping, and the earliest site of ventricular preexitation was found in right anteroseptal/parahisian region, with -38 miliseconds. A 6mm cryocatheter was used for ablation, and just at the 4th second of the first cryomapping, the AP had disappeared (figure1C). Four complete lesions of cryo at -80^{0}C were given on this location. There was no complication during the procedure, but incomplete right bundle branch block. On the 9. month of follow-up, left ventricular functions and dyssyncrony measurements were found totally improved (LV ejection fraction 69%, IVMD=19 ms and SPWMD=5 ms, Figure 1B).
This case is one of the youngest reported infants with successful catheter ablation due to dyssynchrony-related cardiomyopathy. This case also showed that cryoablation can be safely performed in anteroseptal manifest AP even in infants.

P-12
Prevalence of Early Repolarisation Pattern in Children Without Congenital or Acquired Heart Disease

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Objective: We aimed to find prevalance of ER in children, who were referred to Pediatric Cardiology Department unit for miscellaneous reasons, but in whom standart clinical and echocardiographical investigations failed to reveal any structural or acquired cardiac disease.
Methods: Electrocardiography from 1676 successive healthy children from birth to 17 years old were prospectively recorded and analyzed. ER was defined by ≥0.1mV J point elevation in at least two contiguous inferior or lateral ECG leads. The 'notching' or 'slurring' patterns of ER were noted, as well as the maximal J wave amplitude, the rapidly ascending or horizontal/descending pattern of ST segment and the presence of positive or negative T waves in leads showing ER. Age, gender, QTc interval, Sokolow index, heart rate, cardiac symptoms, gestational week and family history of cardiac disease were compared between ER+ and ER- children.
Results: Study population total number is 1676 of 7400 who meets all the inclusion criteria.
Total number of children with ER was 200 and prevelance of ER pattern was 11,9%. Population of study was 909 (%54) male and 767 (%46) female children.ER pattern was present in 87/909 males (9,6%) and 113/767 females (%14,7) (p=0,0013). Mean age of children in all study population was 7,05±5 years, with and without ER pattern was 10,15 ±4,3 vs 6,63 ± 4,91/years (p<0.001). 132,36 and 32 case of all ER (66,18,16%) was detect respectively in inferior, lateral and inferiolateral leads. Horizontal/ descending ST segment and rapidly ascending ST segment after J wave was observed in 17(8,5% of all ER) and 183(91,5% of all ER) cases. Mean J wave amplitude of all cases was 0.14 ±0.049 mV. J wave elevation was ≥0.2 mV in 101 of the 200 cases (50,5%). We don't detect any j wave ≥0.3 mV and negative T wave. ≥ 0,2 mV j wave in adolescence was found to be higher than other periods. 14,5% of all ER cases was type 1 (D1, V4-6 leads) and 85,5% of all ER cases was type 2 (II, III, aVF or/with lateral leads) ER patern. Type 3 ER patern was not detected. Sokolow index was 22,4 ± 9 mm in children with versus 18,2 ± 7,9 mm in children without ER (p <0,0001). Heart rate was 106 ± 31 bpm in children without versus 86 ± 18 bpm with ER (p= 0,026).ER prevalance in children with <100 heart rate, was higher than rest.
Conclusion: ER is present in %11,9 of children of various age and is related to female gender, an older age, a slower heart rate and a higher Sokolow index.

P-13
Influence of ventricular ectopic beats on the systemic ventricular function in patients with congenital heart disease. A long-term longitudinal study.

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Introduction: Isolated premature ventricular contractions (PVCs) in children are generally regarded as a benign phenomenon. Although PVCs have been shown to correlate with impaired cardiac function in adults, this correlation remains controversial in children. The influence of PVCs on systemic ventricular function is of some interest in congenital heart disease (CHD) patients, as these patients are especially prone to ventricular dysfunction. The aim of this study was to evaluate the influence of PVCs on the systemic ventricular function of patients with CHD during long-term follow-up. METHODS: The database of the Heart Center Leipzig was analyzed retrospectively. Key inclusion criteria were: CHD, age 0 – 21 years, initial systemic ventricular ejection fraction (EF) of > 0.35, follow-up of at least 30 months and at least one Holter ECG every year. Patients were classified into 2 groups in accordance with daily PVC burden (Group A > 1% PVCs/ 24 hours, Group B < 1% PVCs/24 hours). A subgroup A1 was defined, which presented a PVC burden of > 1% that persisted from the time of inclusion to the last follow-up. RESULTS: 97 consecutive patients were included with a median follow-up of 84 months (range 33-196). Especially patients of subgroup A1 showed a clinically significant decrease in left ventricular ejection fraction (LVEF, p= 0.03). Possible risk factors like coupling interval or the QRS duration of a PVC did not have any influence on systemic ventricular function. CONCLUSION: The current study underlines a negative influence of PVCs on the systemic ventricular function in patients with CHD during long-term follow-up.

P-14
Is RV resynchronization the key to cardiac remodeling?

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Introduction: Right ventricular dysfunction is an important issue after surgical correction of a tetralogy of fallot. Dysfunction is thought to be due to residual pulmonary stenosis or regurgitation as well as electromechanical dyssynchrony. Currently there are only limited data, particularly focussing on the latter aspect.
Case presentation: A 32-years-old male patient with repaired tetralogy of fallot presented with impairment of the right ventricular (RV) function resulting in a reduced physical capacity (NYHA class II). Cardiac magnetic resonance imaging showed a decreased RV-EF of 31% despite only mild regurgitation and no significant stenosis of the pulmonary xenograft. The ECG showed a cRBBB with a QRS duration > 180 ms. Therefore we opted for a RV cardiac resynchronization therapy (RV-CRT). Two bipolar endocardial leads have been placed in the RV. One at the RV free

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

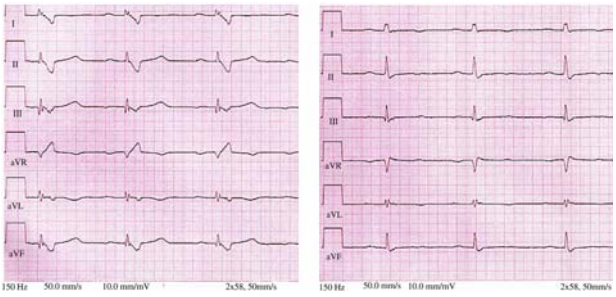

Figure.
Left: ECG (50mm/s) before CRT, right: ECG (50mm/s) after CRT.

wall and a second in the RV apex. An atrial lead was placed in the atrium and the AV delay was set to achieve narrowing of the QRS width via fusion of the paced and intrinsic ventricular depolarization wave.
Results: Post-interventional the QRS duration was markedly decreased and echocardiographically the left ventricular function has improved. Yet, the right ventricular function showed no measureable improvement in the 6-months follow-up, but the MRI displayed a reduced right ventricular enddiastolic volume. Subjectively the physical resilience has remained unchanged.
Conclusions: Studies showed that the possibility of a RV remodeling is diminished, if the QRS length exceeds 150 ms, therefore the possibility in our patient was reduced beforehand. Nevertheless after the electric resynchronization we see a relevant increase in the left ventricular function, supposedly due to a better preload. To evaluate the relevance of RV-CRT in patients with RBBB after cardiac surgery and reduced RV function further studies need to be implemented.

P-15
Prevalence of early repolarization during childhood

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Introduction: Early repolarization (EPR) is considered common in adults and healthy individuals and has been considered as a benign electrocardiographic finding on the 12-lead electrocardiogram (ECG). The prevalence and significance of the ERP in children is unknown. Our study was designed to establish the prevalence of EPR in healthy kids and identify differences between genders.
Methods: We analyzed ECGs of 420 healthy children aged between 1 and 16 years who attended for routine checkup from July 2017 until November 2017. Subjects were excluded if transthoracic echocardiogram revealed structural abnormality. The study population was divided into 3 subgroups of 70 males and 70 females each, according to age (Group A: 1 to 5 years, Group B: 6 to10 years, Group C: 11 to16 years). None of our patients had family history of sudden cardiac death. We defined the gender-specific patterns by: 1) the amplitude of the J point ≥1 mV, 2) the angle between the baseline and the ST-segment (ST angle), 3) upward concave positive T-wave.
Results: The average heart rate and QTc were 99 ±14 beats per minute and 400±12 msec, respectively. EPR was recorded in 225 kids (53%). It was more commonly recorded in the inferior leads (50%), in the lateral 8% and in both leads 22%. EPR was more common in males in group C ($p = 0.002$), and least common in females in group A ($p=0.001$). Incomplete right bundle-branch block (IRBBB) was significantly more common among subjects with EPR (N = 155; 68.8%), compared to those without EPR (N = 49; 25%) (P = 0.002). Differences between men and women decreased as age increased. Heart rate and systolic or diastolic blood pressure did not influence any of the ECG variables analyzed.
Conclusion: ERP is a common phenomenon in healthy adolescents. Sex and age seem to be a factor that may influence the ECG pattern of cardiac repolarization. Longitudinal studies are required to determine whether ERP constitutes a true primary arrhythmic disorder or confers an increased mortality risk.

P-16
Association of QTc interval with coronary involvement in Kawasaki disease.

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Introduction: Kawasaki disease (KD) is a self-limited acute vasculitis affecting small and medium-sized vessels. Diagnosis must be quick, based on clinical criteria and supported by supplementary tests. Up to 20-30% of patients have coronary involvement (ectasia or aneurysms).
EKG is an easy-to-perform and reproducible technique. It has low cost and high availability in any medical center. Up to date, no EKG sign has been associated to coronary involvement. Measurement of QTc interval is the most easy, accessible and widely used method for evaluating ventricular repolarization in children.
Objectives: To describe an association between the presence of coronary involvement of patients with KD and the QTc interval manually measured in the 12-lead surface EKG.
Material and methods: Observational retrospective cross-sectional study of a cohort of Spanish and Japanese patients with KD. The coronary involvement and the QTc interval were evaluated in the EKG performed in the acute period of the disease (up to 6 weeks of illness).
Results: We recruited 180 patients with KD. The median age was 31.1 months [IQ:15.3-48.1], 61.8% males. In our sample, 51/180 (28.3%) patients had coronary involvement. 18.3% (33/180) had ectasia and 10% (18/180) coronary aneurysms. 1.1% (2/180) had giant aneurysms. 95.5% (172/180) of all patients and 98% (50/51) patients with coronary involvement received intravenous immunoglobulin.
QTc interval in V5 and V6 was significantly shorter in patients with coronary involvement (V5: median 378 [IQ:364-395] vs 390 [IQ:371-411] ms, p=0.042; V6: median 377 [IQ:364-392] vs 390 [IQ:371-410] ms, p=0.014). A QTc interval <385 ms in lead V6 was associated with a 2.5-fold increased risk of coronary involvement (OR: 2.5 [CI95%:1.2;5.3], p=0.016).
Conclusions: The QTc interval seems to be a promising marker of coronary involvement in the acute phase of KD. Patients with a QTc interval <385 ms may be susceptible of early, intensive treatment. Prospective studies are needed to validate this hypothesis.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-17

Marfan syndrome (MS): other cardiovascular manifestations rather than aortic root dilatation.

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Introduction: Although aortic root (AoR) dilatation and mitral valve prolapse (MVP) are the most frequent cardiac manifestations of Marfan syndrome (MS), these patients can also develop ventricular arrhythmias and dilated cardiomyopathy. Our aim is to describe the incidence of these findings in our population and see if there is any correlation among them.
Materials: Retrospective study of 57 Marfan paediatric patients (under 18) followed up in our ICC clinic from 2000-2018.
Results: Median age of 10 (IQR 8,15); 56% females. 86% with FBN1 mutation (24,6% de novo). 75,4% on medical treatment (29,8% on losartan, 29,8% on BB and 15,8% on both). 17,5% symptomatic, 4 of them had arrhythmias: 2 with supraventricular tachycardia and another 2 with non-sustained ventricular tachycardia (NSVT). No late gadolinium enhancement was found in any of MRIs performed. There were no sudden deaths.
Echocardiographic findings: 56,1% had AoR dilatation [median 31mm (IQR 27,36)]; 73,7% had mitral regurgitation (MR) (17,5% significant degree) and 52,6% had evidence of MVP. 19,3% of the patients showed LV dilatation, all with normal systolic and diastolic function [median LVEF on MM 64,5% (IQR 61,70%), median S wave on TDI 10cm/seg (IQR 8.3,10)]. There was a significant correlation between the presence of MVP and LV dilatation (p=0.005) but LV dilatation seemed not to be related to the presence of a significant MR (p=0.25). None had significant aortic regurgitation.
Electrocardiographic data: 21% of the patients had baseline repolarization abnormalities mainly in form of inverted/flattened T waves in inferior/lateral leads, which was correlated to the presence of LV dilatation (p=0.03) and MVP (p=0.03). 59,6% had other ECG abnormalities mainly mild intraventricular conduction delay and LVH (15,8%). The presence of arrhythmias didn´t show significant correlation to LV dilatation (p=0.16), MVP (p=0.61) nor repolarization abnormalities (p=0.33).
Conclusion: MS can be associated to many other cardiac comorbidities rather than AoR dilatation. The only factor associated to LV dilatation in our series was the presence of MVP. Patients with a dilated LV didn't show ventricular dysfunction. Although no cases of sudden arrhythmic death, 2 patients had runs of NSVT. Ventricular arrhythmias did not correlate with MVP nor LV dilatation.

P-18

Zero-fluoroscopy cryoablation of atrioventricular nodal reentrant tachycardia in children with congenital heart disease: when everything is a mirror picture

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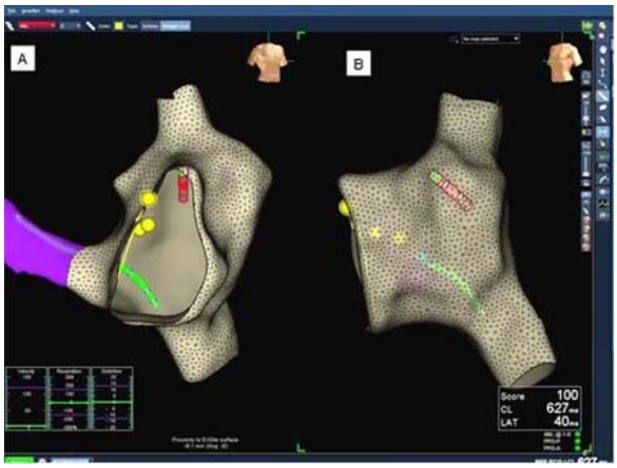
Introduction: Minimizing radiation exposure to pediatric patients during invasive procedures is a critical goal to achieve in Electrophysiology laboratories, as these patients have a high stochastic risk of radiation-induced injuries due to their long life expectancy. In this context, multiple studies support the efficacy and safety of non-fluoroscopic navigation systems to guide ablation procedures in pediatric population. However, there are very few reports on the use of this technique in patients with congenital heart disease.



Methods: We present a six-year-old male with congenital heart disease and episodes of a wide QRS regular tachycardia was scheduled for ablation due to failure of pharmacological treatment. He had recently undergone surgical repair of a double-outlet right ventricle with ventricular septal defect. Besides, he had viscero-atrial situs inversus and dextrocardia.
The procedure was performed under general anesthesia and non-fluoroscopic guidance using the EnSite Precision™ Cardiac Mapping System (St. Jude Medical, Inc.).(figure 1: Right anterior oblique (RAO) and B. left anterior oblique (LAO) view sow situs inversus with dextrocardia: The morphologic right atria is on the left and coronary sinus is on the right. The location of his bundle is in the low medium septum).
Atrial programmed stimulation revealed dual atrioventricular nodal physiology, and reproducibly induced the clinical tachycardia. After AVNRT diagnosis was made, atrioventricular nodal slow pathway ablation was performed.
Cryoenergy was selected for ablation instead of radiofrequency due to these two reasons: 1) The relatively low location of the His bundle in the triangle of Koch, and 2) The higher risk of iatrogenic atrioventricular (AV) block in children with congenital heart disease than in other patients when performing radiofrequency ablation of AVNRT.
Conclusions: In this case, we safely achieved AVNRT ablation using a non-fluoroscopic intracardiac navigation system and cryoenergy ablation.

P-19

Ivabradine for the tratment of automatic atrial tachycardia

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Introduction: Ivabradine is a selective inhibitor of the pacemaker specific current funny. The use of this drug for the control of the heart rate in the adult population, particularly who doesn't tolerate beta blockers therapy, is known. There are, instead,a few scientific papers about using Ivabradine for treatment of automatic atrial tachycardia in pediatric patients.
Methods: We present 5 cases of pediatric patients (age 1-168 months, media age 33,8) who were admitted to our clinic with automatic atrial tachycardia: 2 of them had junctional tachycardia. At admission the mean heart rate of ectopic tachycardia was 160 bpm. No one had heart malformation and ejection fraction was

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

depressed in only one patient with cardiomyopaty and muscolar distrophy.
All patients started beta blockers as first line treatment with poor control of heart rate so in 4 out 5 patients we used association therapy with propafenon, amiodaron or digoxin with only mild control of heart rate.
Results: We use Ivabradine at the dose 0,175 mg/Kg/days till 0,2 mg/Kg/days; in one patient as substitution of beta blocker and in the other in association with digoxin 2 pts, amiodaron and sotalol 1 patient respectively. After introduction of Ivabradine in all we observed recovery of synus rhythm stable in one patient while in the other remain episodes of ectopic tachycardia with a better control of heart rate. The mean heart rate after Ivabradine introduction was of 90-100 bpm. The mean follow up after introduction of Ivabradine was of 9 months (from 1 month till 30 months). We didn't observe acutely or during follow up any side effects.
Conclusions: In accordance to our experience, even if the number of patients treated is small, we can conclude that Ivabradine is usefull for treatment of ectopic atrial-junctional automatic tachycardia not responsive to common therapy. It will be necessary to enlarge number of patients treated with Ivabradine to confirm the usefullness of this drug.

P-20
The Mutations of Genes Associated with Sick Sinus Syndrome in Children

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Sick sinus syndrome (SSS) is life-threatening cardiac arrhythmia, which sometimes can manifest itself with syncope and needs a pacemaker implantation even in children. Sometimes, SSS is accompanied by structural heart diseases such as septal defects, cardiomyopathies, but often the heart is structurally normal. Some mutations of genes associated with high risk of SSS are known. At the same time, the etiology of the syndrome is unidentified and may be genetic caused in 50% of patients with SSS. There are no studies on the prevalence of SSS-associated mutations in children.
The aim of our work is to identify and study the types of mutations of genes associated with high risk of SSS in children.
Methods: We included in the study 19 children (31.5% boys) with severe SSS, from the database of the Russian Pediatric Arrhythmia Center. Personal and family history, physical examination, including ECG, stress test, Holter monitoring, ECHO and other tests, and whole exome sequencing were made. The average age was 8.1±4.5 (from 2 to 17).
Results: In 47% (9 pts) there was the combination of SSS and structural heart disease. 13 children (68%) had syncope, 9 pacemakers were implanted. 7 children (37%) had the mutations of genes associated with SSS: 3 – SCN5A, 2 - HCN4, 1 - TRPM4, 1 - PRDM16. Family history of cardiac diseases was positive in 5 pts; 2 pts had family members with implanted pacemakers.
Conclusion: We found the mutations of genes associated with SSS in 37% of children. Further research and larger patient samples are required to study the prevalence of genetic types of SSS and show the correlation of the genotype with the clinical prognosis.
In addition, our work will enable practitioners to identify children from families with family forms of SSS and sudden cardiac death. Further research can help us determine the criteria for selecting children for genetic testing.

Table.

Parameters, ms	LQTS	Control	P-value
QTc (ECG)	477±51	380±22	0.0000
QTc after TT	492±33	426±17	0.0000
QTc max (HM)	553±43	460±25	0.0000
QTc mean (HM)	496±37	415±21	0.0000

P-21
New Perspectives of Holter Monitoring in Diagnostics of the Long QT Syndrome in the Young

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Introduction: Long QT Syndrome (LQTS) is a hereditary life-threatening cardiac arrhythmia. The prognosis and efficiency of therapy depends on genotype and early diagnosis. About 25-35% of genetically positive LQTS patients have no symptoms with normal QTc values. Additional diagnostic criteria are required to identify silent mutation carriers among apparently healthy. 24-hour ECG Holter monitoring (HM) is used for LQTS to detect ventricular arrhythmias, T-wave alternans and T-wave morphology. The automatic QTc analysis on HM has not been used for diagnostic purposes. We aim at determining the potential significance of HM-based QTc analysis for LQTS diagnosis.
Patients and Methods: 58 children aged 5-17 years with genetically confirmed LQTS (64% boys; 25 patients have syncope, 7 - aborted cardiac arrest, 44 - receive beta-blockers) were randomly selected from 450 LQTS patients (Russian Pediatric Arrhythmia Center database). The age-matched control group consists of 59 children (73% boys) without cardiac pathology. The study protocol includes ECG, treadmill-test (TT), family history analysis. The HM protocol includes QTc max and QTc mean measured automatically and accepted after expert's confirmation.
Results: The mean QTc values for all methods of evaluation were significantly higher in LQTS patients (Table). However, the sensitivity and specificity of methods for the differential diagnostics of LQTS based on an assessment of the QTc were different. In 20% of patients QTc on a standard ECG and in 35% of patients QTc after TT were lower than the once determined by diagnostic criteria (<440 ms and < 480 ms correspondingly). ROC-curve analysis was used for determining QTc cut-points for QTc mean and QTc max categorization. QTc mean > 450 ms and QTc max > 490 ms on HM enabled us to distinguish between LQTS and control in 93% and 100% of cases, and identify 12 (20%) LQTS patients unrevealed by the conventional diagnostic approach.
Conclusions: HM-based QTc criteria are efficient for LQTS diagnostics, especially for the detection of asymptomatic children with "silent" forms of the disease.

P-22
Risk factors for major arrhythmic events in pediatric patients with long QT syndrome

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Introduction/Objectives: Long QT-Syndrome (LQTS) is an inherited arrhythmic disorder associated with sudden cardiac death

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

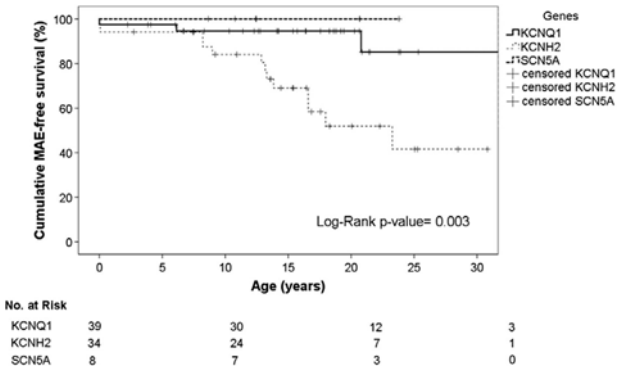

Figure.

(SCD). The goal of this study was to define predictors for major arrhythmic events (MAEs) in pediatric LQTS patients.
Methods: All patients aged ≤ 18 years with a clinical and molecular genetic diagnosis of LQTS type 1, 2 or 3 were included in this retrospective single-center study. Clinical data were recorded by medical chart review. MAEs were defined as the occurrence of SCD, aborted SCD, appropriate implantable cardioverter-defibrillator discharge or sustained ventricular tachycardia.
Results: Childhood onset LQTS was diagnosed in 81 patients (46 males) from 60 families. Median age at diagnosis was 7.6 years (range 0.0 – 18.0 years). A pathogenic or likely pathogenic mutation in the *KCNQ1*, *KCNH2* and *SCN5A* gene was identified in 39, 34 and 8 patients, respectively. MAEs were documented in 16/81 patients (9 males), during a median follow-up time of 5.0 years (range 0.2 – 25.7 years). MAE were more likely in patients carrying a mutation in the *KCNH2* locus (13/16, 81.3%) than in the *KCNQ1* (3/16, 18.8%) or *SCN5A* locus (0/16) (see Figure). QTc-duration was longer in patients with MAE compared to patients without MAEs (570 ± 62 ms versus 497 ± 49 ms, p <0.001, independent t-test). Syncope occurred more often in patients with MAEs (9/16, 56.25%) than in patients without MAEs (11/65, 16.9%) (p= 0.001, Chi-square-test).
Conclusions: Risk factors for life-threatening events included mutations in the *KCNH2* locus, longer QTc-duration and a higher quantity of syncope in this pediatric LQTS cohort.

P-23
Natural history of non-surgical complete atrioventricular block in children and predictors of pacemaker implantation

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Objectives: Data on natural history of non-surgical complete atrioventricular block (CAVB) in children are scarce and criteria for pacemaker implantation (PM) are based on low level of evidence. We aimed to evaluate natural course and predictors of PM in a nation-wide cohort of paediatric patients with CAVB.
Methods: All paediatric patients presenting between 1977 and 2016 with CAVB in absence of any but trivial structural heart disease, were retrospectively identified from the institutional database yielding 95 subjects (female 54, male 41) aged median 4.06 (IQR 0.10-10.34) years at first presentation. Patients were followed-up for median 0.80 (IQR 0.02-7.07) years providing a total of 347 patient-years available for analysis until PM or end of follow-up. Serial 24-hour Holter recordings and echocardiograms were reviewed to asses heart rate profiles, left ventricular (LV) size and shortening fraction. Absolute values and z-scores of healthy population were used for analysis. Predictors of PM performed >1 month after first presentation were evaluated using Cox PH model.
Results: Absolute minimum and mean 24-hour heart rates and maximum RR intervals had a non-linear correlation with age (p<0.0001 for all) with maximum progression of bradycardia during the first 2 years of life. Both LV end-diastolic diameter (median 1.49, IQR 0.27-2.67 z) and shortening fraction (median 36, IQR 31-43%) at presentation were stable throughout follow-up. PM was performed in 64 patients (67.4%) reaching published guidelines criteria. Probability of freedom from PM was 56.9/45.3/42.4/29.7% at 1/3/5/10 years. Mean heart rate at presentation was the strongest predictor of PM (HR=0.939, CI 0.894-0.986, p=0.011 per unit increase) regardless of presentation age. Patients presenting with a mean heart rate >58 BPM (>75 centile of the group) had high probability of freedom from PM within the subsequent 5 years (91.7%) as opposed to rest of the group (41.2%), p=0.012. Echocardiographic parameters did not predict PM.
Conclusions: Paediatric patients with CAVB show an age-dependent decrease in heart rates, stable degree of LV remodelling and preserved LV function. Need for PM can be predicted by the heart rate profile at presentation defining a low risk group and allowing for stratified follow-up.

P-24
Outcome of 3D electroanatomic mapping-guided permanent pacing in parahisian and septal pacing sites in children.

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Introduction: Right ventricular (RV) parahisian, mid septum, outflow tract (PHP-MS-RVOT) pacing sites have been proposed to prevent pacing-induced left-ventricular (LV) dysfunction. 3D-electroanatomic mapping systems (EAM) guide cardiac catheter navigation and reduce fluoroscopy during procedures. The aim of the study was to evaluate the results of EAM-guided pacemaker system implantation (PMI) in PH-MS-RVOT in pediatric patients.
Methods: Children/adolescents with complete atrioventricular block (CAVB) and no other congenital heart defects prospectively underwent EAM-guided PMI into PH-MS-RVOT. With EAM, a geometric reconstruction of right heart was initially performed; then a pacing map identified RV septal sites with narrower paced QRS complex. The maps obtained guided the implantation of atrial/ventricular leads. ECG, LV contractility (ejection fraction, EF, Global Longitudinal Strain, GLS) and synchrony (Systolic Dissynchrony Index, SDI, Septal to Posterior Wall Motion Delay, SPWMD, Interventricular Delay, IVMD) were evaluated before and after implantation, up to 2 years. Data are reported as mean ±SD. *P<0.05 was significant*.
Results: Twenty patients (15 females), 9.9±3.9 years, 35±16 kg, 138±23 cm, underwent PMI (13 VVIR, 7 DDD) in PHP-MS-RVOT (respectively, 7-11-2 patients). Procedure time (167±38 minutes) and radiation exposure (4.8±3.1 mGy, 114±83 microGy/m2) were recorded. DDD patients were older than VVIR (14±2 years vs. 7±2, P<0.001), *had longer procedure time*

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Table 1

	QRS ms	EF %	SDI ms	SPWMD ms	IVD ms	GLS %
Pre-implant	89±23*	64±11	/	/	/	/
Post-implant	108±12* P=0.004	61±6	2.9±2.2	90±24	18±13	-23 ± 3
1 Year	/	62±7	1.7±0.7	87±24	23±8	-23 ± 3
2 Year	/	60±6	2.7±1.1	92±24	20±15	-23 ± 2

(200±35 min. vs. 150±26, P=0.002) and exposure (7±4 mGy vs. 3.6±2.0, P=0.015). One ventricular lead dislodged and was repositioned (censored). QRS duration increased significantly after implantation, LV EF and synchrony were normal throughout follow-up without significant variations (table 1), pacing was 100%. No significant differences were observed between patients with DDD-VVIR, and PHP-MS-RVOT.
Conclusions: EAM-guided PMI in PH-MS-RVOT in pediatric patients showed preserved LV function at 2 years follow-up.

P-25
Single Centre preliminary results of laser lead extraction in paediatric and CHD patients: how multidisciplinary cooperation can make the procedure safe and effective.

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Background: Over the past decades, the number of transvenous pacemaker (PM) and defibrillator (ICD) implantations in paediatric patients markedly increased. Therefore, the amount of children and young patients needing lead extraction is expected to grow. Herein our preliminary experience with laser transvenous lead extraction in children/young CHD patients is reported.
Methods: All patients underwent pre-procedural transthoracic and transesophageal (TEE) 2D-/3D-echocardiography, and iodine contrast Computed Tomography (CT) to assess lead location and course, presence of calcifications, and relationship with venous and cardiac structures in order to evaluate risks and allow appropriate procedural planning. Intraprocedural 3D-TEE was also used both to evaluate lead embedding into venous structures and to real-time monitor integrity of vascular and cardiac structures. All procedures were performed in the hybrid operating room by a multidisciplinary team including electrophysiologists, cardiac surgeons, anesthesiologists, interventional cardiologists (when required), radiology and perfusion technicians. Post-procedural management included at least 24hrs of patient monitoring in intensive care unit.
Results: From 09/2017 to 09/2018, 8 patients (4 females) with a median age of 12 years (range: 11-28) underwent lead extraction. Six had a structurally normal heart, 3 with congenital Complete Atrio-Ventricular Block (cAVB), 2 with cAVB in the context of a myopathy (1 Emery-Dreifuss and 1 Kearns-Sayre Syndrome), and 1 with a channellopathy. Two had Transposition of the Great Arteries (TGA) corrected with a Mustard operation, Sinus Node Disease and obstruction of the Superior Vena Cava (SVC) baffle. One of the TGA patients had a dual-chamber PM, whereas the other 7 had a single ventricular system, 6 PM and one ICD. The indication consisted in lead dysfunction or need for SVC baffle stenting in the post-Mustard patients.
Median age of the 9 removed leads was 4.5 years (range: 1.5-16). Electrodes were extracted through a left subclavian venous approach using 40-80 Hz laser sheaths in all, alternated with mechanical sheaths in the two cases with previous Mustard. No complications occurred during/after extraction.
Conclusions: The use of pre- and intra-procedural imaging and a multidisciplinary approach may ensure successful and safe outcome of laser lead extraction even in young patients with relatively old leads and/or corrected CHD.

P-26
The approach to drug therapy of arrythmias in children with suspected myocarditis

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Objective: To analyze the antiarrhythmic therapy (AT) effectiveness of ventricular (VA) and supraventricular arrythmias (SVA), which first were registered in children with suspected myocarditis.
Materials and Methods: We have analyzed the data of 40 patients, mean age 5,7±5,5 years (from 12 months to 17 years): 21/40 with VA and 19/40 with SVA. The disease duration before the first examination in our clinic was 20.2±39.2 months (0-104 months). We used all groups of antiarrhythmic drugs in recommended dosages according to age and anthropometric parameters of patients. Efficiency control was carried out by Holter monitoring with following criteria: reducing the number of single premature beats (PB) by more than 50%, double PB by at least 90% and the absence of ventricular/supraventricular tachycardia. In cases of ineffectiveness, we have varied prescribed medication to another after 5 half-lives periods or used combined AT.
Results: The follow-up period was 21.2±17.98 months (2-66 months). AT was prescribed to 95%(38/40) patients (20/38-VA; 18/38-SVA). Patients received from 1 to 7 drugs consistently, combined AT was required in 34.2% (4 with VA and 9 with SVA).
In VA group β-blockers was prescribed in 85%(17/20): Propranolol in 60%(12/20), Metoprolol tartrate in 20%(4/20), Nadolol in 5%(1/20). Amiodarone was used in 70%(14/20) cases. IC class - in 66,6%(12/20): Propafenone in 55%(11/20), Flecainide in 15% (3/20), Lappaconitine hydrobromide in 10%(2/20). Also Sotalol (1/20) and Verapamil(1/20) were used. AT was effective in 55%(11/20) cases, partial efficiency was detected in 20%(4/20), lack of effect –25%(5/20), and in one of them negative result with the development of ventricular fibrillation was observed.
In SVA group β-blockers was prescribed in 86,9%(16/18): Propranolol in 44,4%(8/18), Metoprolol tartrate in 44,4%(8/18). Amiodarone was used in 55,6%(10/18) cases. IC class - in 55,6% (10/18): Propafenone in 38,8% (7/18), Lappaconitine hydrobromide in 16,6%(3/18). Also Digoxine(3/18), Sotalol(1/18) and Ivabradine(1/18) were prescribed. The treatment was effective in 61,1%(11/18) and ineffective in 38,9%(7/18) cases. SVAs weren't observed after discontinuation of AT in 6 patients (33,3%).
Conclusion: The choice of AT for children with suspected myocarditis was strictly individual. B-blockers in most cases were the first line of therapy. In both groups we often observed insufficient result of AT.

P-27
Radiofrequency ablation of atrial arrhythmias in children: one-center experience

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Objective: To analyze the radiofrequency ablation (RFA) results of atrial arrhythmias (AA) in children
Materials and Methods: The study group included 45 patients with AA: chronic focal atrial tachycardias (ATs) - 8(17.8%), recurrent focal ATs - 15(33.3%), paroxysmal ATs - 14(31.1%), frequent atrial premature beats - 2(4.4%), atrial flutter (AF) - 4(8.9%), atrial fibrillation (AFib) - 2(4.4%). There were 29 (64.4%) boys. The mean age was 10.5±6.5 (3,6-17,9 y.o.). Age of AA debut was from 2 months to 17 years (6.2±4.7). The etiology: idiopathic in 68,9% (31/45), suspected myocarditis in 22,2%(10/45), post-incisional in 8,9%(4/45), CHD in 2,2%(1/45). Arrhythmia-induced cardiomyopathy was established in 8,8% cases. All patients received antiarrhythmic therapy prior to RFA.
Results: Between 2008 and 2017, 45 patients underwent 52 RFA. One-year follow up was possible for 40 patients (88,8%). The mean age of primary RFA was 14.3±2.75 y.o. (from 3.6 to 17). Substratum localization: 6-crista terminalis, 5-coronary sinus ostium, 5-auricle of the LA, 3-anterolateral wall of the RA, 2-posterior-lateral wall of the RA, 1-near to AV node, 1-auricle of the RA, 3-interatrial septum, 5-cavatricuspid isthmus, 2-pulmonary veins, and in 12 cases AA was multifocal. The primary RFA efficiency was 77.8%(35/45), the second - 84.4%(38/45). The ineffectiveness was most often associated with complexity of arrhythmia's induction and mapping or in multifocal AT. We observed early AA recurrence (within 7 days) in 9 cases (17.3%) and in 3 cases (5.8%) - after 4, 7 and 12 months. Arrhythmia recurrence wasn't associated with substratum localization or etiology. AFib was inducted intraoperatively in 15.3%, AF with transition to AFib in 5.8%, AF+AFib in 3.9% in the cases of right AA only. Electrical cardioversion was carried out in 9.8% cases. Subtotal AV block developed in one patient.
EMB were performed in 7 patients: chronic myocarditis –4/7, cardiomyopathy –1/7, no pathology –2/7.
Conclusion: The primary RFA efficiency was 77.8%, the second -84.4%. The most difficulties were in cases of multifocal AT, non-sustaned AA during EP study and the younger age group. In our study intraoperative induction of AFib/AF was observed only in patients with right-located AAs. One-year follow up shows good results in 70% patients.

P-28
Outcome of arrhythmias in children with suspected myocarditis

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Aim: The aim of this study was to analyze the clinical characteristics and outcome of arrhythmias in children with suspected myocarditis.
Materials and methods: 52 patients with arrhythmias and suspected myocarditis were included, who were examined in our center in the period from 2011 to 2018. Mean age 5,7±5,5 years (from 12 months to 17 years).
The criteria for inclusion: the presence of arrhythmia, suspected myocarditis, at least two examinations in the dynamics. 21 patients had ventricular arrhythmias, 19 - supraventricular arrhythmias, 10 - high degree atrioventricular block (AVB) and sinus node dysfunction-2.
Patients underwent a complete history, physical examination, laboratory studies (including thyroid function, CK, CK-MB, LDG, Troponin I, proBNP, serology studies), chest X-rays, echocardiography, ECG, Treadmill test and Holter monitoring (HM). Cardiac MRI and Endomyocardial Biopsy (EMB) were performed according to indications and physician's decision.
The diagnosis of the myocarditis was made in the presence of association between first appear arrhythmias and viral infection and in combination with: elevated markers of myocardial damage and/or cardiomegaly by chest radiography and/or increased left ventricular end diastolic and systolic dimensions with/without decreased ejection fraction.
Results: duration of follow-up was 22,05±18,7 months (6-82) months.
All patients received myocarditis therapy, antiarrhythmic therapy received 38 patients (20 with VA; 18 with SVA).
In the group of patients with ventricular arrhythmias, normalization of rhythm was observed in 10/20 patients; reduction of the frequency and duration of episodes of arrhythmia and rhythm control is on the background of antiarrhythmic therapy – 5/20; lack of antiarrhythmic effect – 5/20, RFA was performed in 4 patients of 5.
In the group of supraventricular arrhythmias normalization of rhythm – 11/18 children, reduction of the frequency and duration of episodes of arrhythmia and rhythm control is on the background of antiarrhythmic therapy – 7/18; RFA was performed in 3 patients of 8.
In the group of patients with AV and sinus node dysfunction - the changes were irreversible in all children. A permanent pacemaker was implanted in 10 patients.
Conclusion: in our study in children with suspected myocarditis, tachyarrhythmias in most cases were reversible in contrast to bradyarrhythmias.

P-29
Asymptomatic WPW pattern in children: a single center experience with the use of isoproterenol in the arrhythmic risk stratification

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Introduction: Lethal arrhythmias and sudden death are reported in asymptomatic children with a Wolff-Parkinson-White(WPW) pattern. Electrophysiologic study(EPS) has been advocated in order to identify high risk patients that may benefit from prophylactic ablation of the accessory pathway(AP). We aimed to report a single centre experience with risk stratification in asymptomatic children with WPW pattern using EPS.
Methods: We retrospectively analyzed records of asymptomatic patients with WPW pattern who underwent EPS(transesophageal or endocavitary) at our Institution since 2012. Parameters evaluated were: AP antegrade effective refractory period(aERP), shortest cycle length(1:1 SCL)sustaining 1:1 conduction over the AP, inducibility of atrio-ventricular re-entrant tachycardia(AVRT) and/or atrial fibrillation(AF) and shortest preexcited R-R interval (SPERRI) during AF. Parameters were determined at baseline and after iv infusion of isoproterenol. Patients were considered at risk and therefore proposed for AP ablation in case of AVRT induction with at least one of the following risk factors: aERP≤250 msec, 1:1SCL≤250msec or AF induction with a SPERRI≤250 msec. Other indications for ablation were sport eligibility, parental choice and significant ventricular dissynchrony.
Results: Out of forty-seven patients(11±2 years,29 males), 5(10%) showed decremental properties suggesting Mahaim-type

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Table 1

AVRT baseline	AVRT isoproterenol	aERP baseline	aERP isoproterenol	Mann-Withney-Wilcoxon	1:1SCL baseline	1:1SCL isoproterenol	Mann-Withney-Wilcoxon	AF/flutter baseline	AF/flutter isoproterenol
12pts (29%)	17pts (40%)Relative risk+25% (no statistical difference)	314±50 msec	250±27 msec	p<0.0001	308±73 msec	226±32 msec	p<0.0001	9pts (21%)	6pts (14%)

preexcitation. For the other 42, EPS results are reported in Table1. Patients at risk were 16/42(38%). Considering only AP properties, 14(33%) had aERP/1:1SCL≤250 msec at baseline and additional 13(30%) during isoproterenol. Only 3 patients(7%) fulfilled all risk criteria. A total of 24 patients(57%) underwent ablation(no procedural complications), which was acutely successful in 23(95%). Three recurrences occurred, 2 of whom underwent subsequent successful ablation. The remaining 18 patients(43%) are still asymptomatic (3-year mean follow-up).
Conclusions: In asymptomatic children with WPW pattern, isoproterenol during EPS induced significant reduction of aERP and 1:1SCL and increased the cases of inducible AVRT. In our experience the use of isoproterenol increased the number of patients considered to have potential high risk for lethal arrhythmias that could benefit from AP ablation.

P-30
Increased heart rate variability in children with Fontan circulation and sinus node dysfunction

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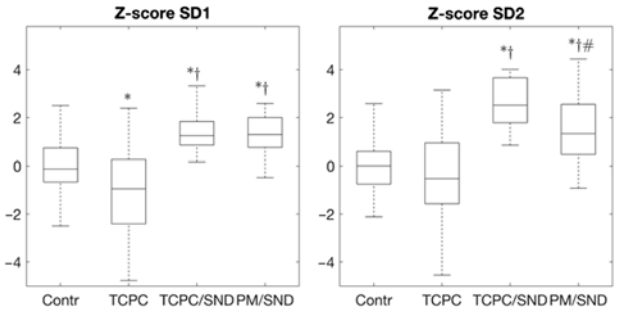
Introduction: Development of sinus node dysfunction (SND) causes significant morbidity in patients who have undergone Fontan surgery. Heart rate variability (HRV) is a marker of the autonomic nervous system regulation of the heart. HRV has been shown to be reduced in patients with Fontan circulation. Changes in HRV have been associated with tachyarrhythmia as well as SND in adult patients. We aimed to study whether changes in HRV could be detected in 24-hour electrocardiographic (ECG) recordings in Fontan patients with SND.
Methods: HRV from 24-hour ECGs was analyzed by Poincaré analysis where SD1 represents the magnitude of the beat-to-beat variability, and SD2 represents changes in mean RR. We compared HRV parameters using analysis of variance with groupwise t-tests as post-hoc tests: patients with Fontan circulation who later developed the need for a pacemaker due to severe SND (n=12), patients with Fontan circulation and SND but without indication for pacemaker treatment (n=11), patients with Fontan circulation without SND (n=90) and healthy controls (n=66).
Results: Patients with Fontan circulation without SND had significantly lower SD1 than controls. Both SD1 and SD2 were significantly higher in the two SND groups compared with both healthy controls and patients with Fontan circulation without SND. SD2 was slightly reduced in SND patients with pacemaker compared to SND patients without pacemaker (p=0.06).



Conclusions: As expected, patients with Fontan circulation showed lower HRV than healthy controls. When the patients develop SND the HRV increases and becomes significantly higher compared to healthy controls and Fontan patients without SND. However, SD2 tended to decrease again in SND patients with pacemaker, which could indicate a reduced diurnal variability in heart rate. This was a small study, but our results indicate that HRV analysis might be useful in the follow-up of Fontan patients regarding development of SND.
Poincaré indices presented as z-scores based on controls. Boxes represent median and interquartile range, whiskers show range. P-values were derived from post-hoc tests after analysis of variance. * p<0.05 versus controls. † p<0.05 versus TCPC. # p=0.06 TCPC/SND vs TCPC/PM /SND.
TCPC=total cavopulmonary connection, TCPC/SND= TCPC with SND, PM/SND=TCPC with SND and pacemaker.

P-32
Videoscopic cardiac sympathetic denervation for pediatric patients – one center experience

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Introduction: Left or bilateral cardiac sympathetic denervation (CSD) in patients with life-threatening ventricular arrhythmias (VA) may be supportive therapy. Recently thoracoscopic cardiac sympatectomy as minimally invasive method is used. We present our experience in the method in pediatric patients.
Material and methods: During last ten years video-assisted thoracoscopic CSD was performed in 16 patients (8 boys) age from 6 weeks to 17,8 years. Thirteen children suffered from life-threatening VA, 11 pts were after aborted cardiac arrest, 8 had syncope attacks. In 11 of them long QT syndrome (LQTS) was diagnosed, in 2 catecholaminergic polimorphic ventricular tachycardia, in 1 familiar short QT syndrome, 1 girl had hypertrophic and 1 dilated cardiomyopathy. Before CSD all patients were on propranolol and 6 additional on mexiletine. In 13 pts ICD was implanted, in 9 before CSD – 6 of them had appropriate interventions. On standard ECG mean QT/QTc interval (according to Bazett's formula) was 479/520 ms.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Result: CSD (left in 15 and bilateral in 1 patient) was successfully performed under general anesthesia. In two patients pleural effusion accumulated early after the CSD procedure. Mean QT/QTc interval on ECG performed before discharge from the hospital was 468/504 ms. Follow-up period lasting from 2 months to 4,3 years (mean 2,9 years). At the time 13 of our patients with ICD had no appropriate interventions. The girl with hypertrophic cardiomyopathy had a heart transplant. In one boy appropriate ICD interventions appeared 3 years post left CSD despite continuous therapy of propranolol and mexiletine in high doses, he died 4 months later during electrical storm. Another boy with extremely long QT/QTc interval died seven months after bilateral CSD also during electrical storm. We did not observed serious side effects associated with the CSD procedure.
Conclusions: Videoscopic cardiac sympathetic denervation is a minimally invasive method of treatment and can be performed even in the youngest patients. QT/QTc intervals shortened slightly after CSD, the number of adequate ICD interventions decreased. CSD seems to be a good additional method of therapy for the majority of pediatric patients with life-threatening VA, especially with LQTS but further research is needed.°

P-33

Long-term Follow-up after epicardial Pacemaker Implantation in Neonates and Infants: a single center experience

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Introduction: A variety of different surgical techniques and approaches in pacemaker (PM) implantation within the first year of life are used depending on the implanting center's preference. At our institution, we advocate the surgical implantation of epicardial leads to avoid problems associated with transvenous electrodes in a growing child. Our aim is to identify possible long-term benefits and disadvantages of our approach in PM implantation in neonates and infants.
Method: With this retrospective study, we looked at patients undergoing PM implantation within the first year of life at our center. Atrial and ventricular lead sensing and capture thresholds at implantation, after 1, 3, 5 and 7 years and maximal follow-up time in each patient were analyzed. Subgroup analysis was performed in acquired versus congenital atrioventricular block, implantation below or above 1 month of age and with or without previous heart surgery.
Results: A total of 52 consecutive patients at a median age at implantation of 3 (0 – 10) months were identified. PM indications were postoperative atrioventricular block (n = 33), congenital atrioventricular block (n = 12) and sinus node dysfunction (n = 3). During a median follow-up time of 40.4 (range: 0.1 – 114) months median sensing remained between 3.1 and 4.0 mV for atrial leads and between 10.0 and 14.4 mV for ventricular leads. Pacing thresholds were 0.7 V for atrial leads and 1.2 V for ventricular leads. There was no adverse pacing effect on left ventricular function and dimensions over time. 20 PM related reoperations had to be performed in 13 / 52 (= 25%) patients. Indications for these reoperations consisted of infection (n=3), battery exhaustion (n=10), generator dislocation (n=3), lead dysfunction (n=3) and diaphragmatic paresis (n=1). There was no pacemaker-related mortality. No significant differences in ventricular pacing threshold in various etiologies were found. Median interval from implantation to first generator (and/or electrode) replacement was 44 (0.7 – 98) months.
Conclusion: Our results show that epicardial PM implantation in neonates and infants is a safe and effective procedure. Our current implant technique and pacing strategy shows good long- and mid-term results with a low rate of complications.

P-34

Cardiac involvement in Noonan syndrome: our experience

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Introduction: Noonan syndrome (NS) is an autosomal dominant disorder with a high incidence of cardiovascular disease. PTPN11, RAF1 and SOS1 are some of the mutations that correlate most with cardiac pathology. Pulmonary valve stenosis (PVS) and hypertrophic cardiomyopathy (HCM) are the most frequently associated. HCM can represent a risk factor for arrhythmias or sudden death. Our objective is to study the prevalence of heart disease in our patients with NS.
Methods: We reviewed the patients diagnosed with NS at our center in the last 30 years and analyzed the cardiac involvement data collected in the electronic medical record.
Results: We obtained a sample of 41 subjects, of which the majority were male (56,1%). Only 9,7% of the subjects did not have heart disease. Among the congenital heart defects detected, 56,1% were PVS, followed by HCM 24,4%, 50% with biventricular hypertrophy). Other pathologies detected were: atrial septal defect 'ostium secundum' type (17%) and atrioventricular septal defect (4,9%), aortic coarctation (4,9%), dysplastic mitral valve (4,9%), idiopathic dilatation of main pulmonary artery and branches (7,3%). Less frequent (2,4% each) were: pulmonary atresia, tetralogy of Fallot, pulmonary arterial hypertension (PAH) and ductus. Regarding the need for some type of interventionism: 14 subjects (34,1%) required cardiac surgery and 8 (19,5%) underwent catheterization for the ones with PVS. Four patients were under treatment, mainly beta-blockers. Regarding electrocardiographic findings: 26,8% had a normal ECG, with the remaining presenting changes in the axis, with a greater percentage of left deviation (19,5%) and left anterior fascicular block in 14,6%. Two subjects had arrhythmias which consisted of frequent premature atrial beats. PTPN11 was the most frequent mutation (19,5% of all records) with PVS as associated heart disease, followed by RAF1 that was associated more with HCM. There were two deaths secondary to respiratory infection.
Conclusions: Our findings are quite similar to what is described in the literature. PVS remains the most frequent finding followed by HCM. These patients also have a very typical electrocardiographic pattern and a high rate of interventionism. None ventricular arrhythmias nor sudden death were documented.

P-35

Hypertrophic Cardiomyopathy, Catecholaminergic Polymorphic Ventricular Tachycardia. Does genetics always help?

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Introduction: Hypertrophic cardiomyopathy (HCM) is a common genetic cardiovascular disease (ie, 1/500) and is a major cause of

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

sudden death. Nowadays, in more than 60% of cases is possible to identify a genetic mutation and the vast majority of genes responsible for HCM encode proteins associated with the sarcomere.
Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an hereditary disease characterized by the development of adrenergically-mediated ventricular tachycardia in individuals with an apparently normal heart. There are described mutations in 6 genes, most of them are caused by the mutations in the cardiac ryanodine receptor gene RyR2.
Mutations in Ca2+-handling proteins can contribute to the pathogenesis of HCM. We identify a mutation in RYR2 gene in a patient with HCM phenotype.
Clinical Case: Our patient is a 12 years old girl checked for the first time for nonspecific chest pain.
Family past history negative for cardiac problems.
Physical examination and baseline ECG were normal
An echocardiogram revealed a maximum left ventricular wall thickness of 20 mm at the basal segment without left ventricular outflow obstruction. This hypertrophic zone had an increase echogenicity appearing like brightness cardiac mass. The left ventricular function was normal
Subsequently the patient underwent a cardiac MRI that revealed hypertrophy of the basal segment with helicoidal appearance, with increase of signal intensity in T1 and T2 indicative of fibrosis and revealed late gadolinium enhancement in this area compatible with a patchy fibrosis.
Stress test was normal, neither isolated premature ventricular contractions nor VT was induced. 24-hour Holter monitoring was normal as well.
Full panel testing was ordered for HCM finding a mutation in RYR2 p.332R>W, possibly pathogenic. This variant has been associated with CPVT.
The family screening in her parents and her brother was normal
The patient was placed on beta-blockers without seeing arrhythmic events during follow-up
Conclusion: We describe a case of a patient with HCM with a genetic mutation in RYR2 p.332R>W. This variant is associated with CPVT and it is not found in general population.
Although RYR2 has been associated with HCM, the molecular and cellular mechanisms that link a RyR2 mutation with the development of HCM are completely unknown. Perhaps mutations in the ryanodine gene, that result in a dysfunctional release of calcium, lead to pathological cardiac remodeling.

P-36
The variable correlation between genotypes and phenotypes in cardiomyopathies and arrhythmogenic syndromes

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Introduction: Cardiomyopathies and arrhythmogenic syndromes have a considerable variability in genotypes and phenotypes. Modifier genes, epigenetic and environmental factors have been suggested as main determinants for different phenotypes associated with the same genotype.
Methods: We carried out Next Generation Sequencing (NGS) analysis on PGM-Ion Torrent platform in 9 probands with cardiomyopathies or arrhythmic syndromes, using a panel targeted approach which included 21 disease-causing genes. A Sanger sequencing was taken to confirm the results and a co- segregation analysis was performed in family members.
Case series examples: Case 1. In a 9-years-old female with QT-long syndrome, molecular testing detected a de novo likely pathogenic variant in KCNH2 (c.205C>G;p.Leu69Val) and 3 variants of uncertain significance (VUS) in DSP (c.5056C>T;p.Gln1686Ter), MYH6 (c.5465G>A;p.Arg1822Gln) and CAV3 (c.387C>A;p.Cys129Ter), all found in her mother affected by Dilated Cardiomyopathy (DCM). Case 2. A 6-years-old male with hypertrophic obstructive cardiomyopathy (HOCM), presented a VUS in MYBPC3. This was also found in his mother, who did not suffer of any cardiac disorder. Case 3. In a 1-year-old male with diagnosis of DCM, a likely pathogenic variant of MYH7 and a VUS of RYR2 were detected. The first variant was found in his father and the second in his mother. Although carriers both parents were phenotypically healthy.
Discussion: Our case series confirm that: 1. A single gene mutation often is not sufficient to bring a cardiac disease. 2. The same gene mutation can produce different types of diseases. 3. Some genetic heart disorders come from the combination of multiple gene mutations. The relationship between genotype and phenotype is made even more complex by incomplete penetrance and variable expressivity of some genes.
Conclusion: NGS-based analysis has an high diagnostic potential, since it allows to identify gene mutations that are likely related to genetic heart disease. Pooled data from collaborative studies and larger case series are needed to define more accurately the complex relation between genotype and phenotype in cardiomyopathies or arrhythmogenic syndromes.

P-37
The Relationship between Plasma Fibroblast Growth Factor-23 Concentration and Preclinical Cardiovascular Damages in Children with Primary Hypertension

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Objective: Fibroblast growth factor-23(FGF-23) has been reported to be associated with cardiovascular diseases including hypertension. This study is to investigate the association between plasma FGF-23 and preclinical cardiovascular damages.
Methods: With prospective study, 77 patients with primary hypertension were enrolled. The 2004 NHBPEP diagnosis criteria were referred. Basic clinical features were recorded. Carotid wall intima-media thickness (cIMT) and left ventricular hypertrophy (LVH) were assessed as index of preclinical cardiovascular damages. Patients were divided into increased cIMT group and normal cIMT group. LVH and left ventricular geometry was identified, and patients were divided into normal geometry, eccentric remodeling, concentric remodeling and concentric increased LV mass group. Concentration of plasma FGF-23 was detected in all children by ELISA test, and differences were analyzed.
Results: There were 27 patients (35.1%) with LVH in the whole 77 patients, while 50 patients (64.9%) without LVH. There were 64 patients who had cIMT data, while 18 patients (28.1%) with increased cIMT and 46 (71.9%) with normal cIMT. The concentration of plasma FGF-23 in children with increased cIMT (n=18) was higher than normal group(n=46) [55.6(46.2, 63.5) RU/ml vs 48.6(39.4, 57.3) RU/ml], which showed the positive relationship between plasma FGF-23 and cIMT (r=0.222, P=0.032). ROC curve analysis showed the cutoff value was 53.9RU/ml with the predictive sensitivity of 55.6% and the specificity of 71.7%. The concentration of plasma FGF-23 was significantly higher in

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

patients with LVH than those with normal geometry [55.0(46.8, 65.7) vs 48.2(39.5, 56.0)], which showed the positive relationship between plasma FGF-23 and LVH(r=0.224, P=0.018). The concentration of plasma FGF-23 in patients with concentric remodeling (n=10) was significantly higher than that of the normal geometry (n=50) (P=0.036). ROC curve analysis showed the cutoff value was 49.1 RU/ml with the predictive sensitivity of 70.4% and the specificity of 60.0%.
Conclusions: There were some patients who had preclinical cardiovascular damages including LVH and increased cIMT at diagnosis. The concentration of plasma FGF-23 in children with primary hypertension was positively related with LVH and cIMT, and it has certain diagnosis predictive value for preclinical cardiovascular damages.

P-38
Altered methylation levels of imprinting genes in children with congenital heart disease

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Abstract
Background: Congenital heart disease (CHD) is likely resulted from both genetic aberration and environmental adverse factors. Imprinting genes, which are regulated by epigenetic modifications, are essential for the normal embryonic development. However, the epigenetic role of imprinting genes in the pathogenesis of CHD remains unclear.
Objective: To examine the methylation modifications of differences imprinting genes in CHD in order to explore potential epigenetic pathogenesis of CHD.
Design: Eighteen imprinting genes associated with early embryonic development were selected to compare the methylation alternations between 27 children with CHD (ventricular septal defect in 17, atrial septal defect in 12, valvular heart disease in 7, tetrology of Fallot in 4, patent ductus arteriosus in 4, constriction of aortic arch in 3) and 28 healthy controls by using MassArray platform.
Results: The methylation levels of 8 imprinting genes were significantly different between CHD group and Control group. Among them, the methylation levels of imprinting genes GRB10 (51.12% VS 43.42%, P<0.001) and MEST (56.60% VS 53.22%, p=0.025) were significantly higher in CHD group, while those of imprinting genes INPP5F (67.23% VS 73.17%, P<0.001), PEG10(45.17% VS 50.92%, P=0.002), NAP1L5(62.12% VS 68.86%, P=0.007), PLAGL1(40.97% VS 42.80%,P=0.018), NESP(31.31% VS 41.12%, P<0.001) and MEG3(39.53% VS 45.31%,P<0.001) were significantly lower in CHD group. However, no significant imprinting modification changes were found among different types of CHD.
Conclusions: The altered methylation levels of imprinting genes found in our study may imply an epigenetic multi-misregulation in the pathogenesis of CHD during embryogenesis. Further studies are warranted to examine potential variations in these imprinting genes in the pathogenesis of CHD.

Key words: Congenital heart disease, Imprinting, gene, Methylation

P-39
Evaluation of the mean platelet volume and neutrophil-to-lymphocyte count ratio in patients with Kawasaki Disease

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Objective: Kawasaki disease (KD) is considered a kind of systemic vasculitis that primarily invades the medium-sized muscular arteries, including coronary arteries. Platelets are essential players in thromboembolic and inflammatory mechanisms. Mean platelet volume (MPV) and platelet distribution width (PDW) are correlated with platelet function and may be a more sensitive index than platelet number as a marker of clinical interest in various disorders. Neutrophil-to-lymphocyte (N/L) ratio is a predictor of inflammation and it has been shown that associated with cardiovascular events. The aim of this study was to investigate the alterations in MPV, PDW, and N/L ratio in children with KD.
Methods: The study population consisted with 24 KD patients and 20 healthy controls. In all subjects, complete blood count, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR) were measured at the first and tenth days of the diagnosis and compared with the healthy subjects.
Results: There was no significantly difference between the children with KD and control groups for the sex and age (p>0.05). Eight patients were incomplete Kawasaki disease. Cardiac involvement, including pericarditis, endocarditis and coronary artery lesion was detected in the twelve patients. Among these patients, eight had coronary artery involvement. ESR, CRP, WBC, and N/L ratio were significantly higher in the patients at the 1st day compared with the 10th day and control group (p<0.001). N/L ratio was found as 4.2±2.6 / 0.97±0.84 / 0.94±0.56 at the 1st and 10th days and control group respectively. Platelet count and MPV were higher at the 1st and 10th days compared with control group (p<0.05). MPV was found as 7.54±1.1 / 7.7±1.3 / 6.6±0.87 fL at the 1st and 10th days and control group respectively. There was no significantly difference between these groups for the PDW (p>0.05).
Conclusions: Increased MPV and N/L ratio were detected in patients with KD. The present findings emphasized the association between that MPV, N/L ratio and KD. MPV and N/L ratio may be used to determine activity of KD, as a new biomarker. Further studies in larger series including patients with coronary artery aneurysm should be performed in this issue.

P-40
Natural and Undetermined Sudden Death: Value of Post-mortem Genetic Investigation

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Background: Sudden unexplained death may be the first manifestation of an unknown inherited cardiac disease. Current genetic technologies may enable the unraveling of an etiology and the identification of relatives at risk.
Objective: The aim of our study was to define the etiology of natural deaths, younger than 50 years of age, and to investigate whether genetic defects associated with cardiac diseases could provide a potential etiology for the unexplained cases.
Methods: Our cohort included a total of 789 consecutive cases (77.19% males) <50 years old (average 38.6±12.2 years old) who died suddenly from non-violent causes. A comprehensive autopsy was performed according to current forensic guidelines. During autopsy a cause of death was identified in most cases (81.1%), mainly due to cardiac alterations (56.87%). In unexplained cases, genetic analysis of the main genes associated with sudden cardiac death was performed

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

using Next Generation Sequencing technology. Genetic analysis was performed in suspected inherited diseases (cardiomyopathy) and in unexplained death, with identification of potentially pathogenic variants in nearly 50% and 40% of samples, respectively.
Conclusions: Cardiac disease is the most important cause of sudden death, especially after the age of 40. Close to 10% of cases may remain unexplained after a complete autopsy investigation. Molecular autopsy may provide an explanation for a significant part of these unexplained cases. Identification of genetic variations enables genetic counseling and undertaking of preventive measures in relatives at risk.

P-41
Predictors of imunoglobulin resistance and cardiac complications in kawasaki disease

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Kawasaki's disease (KD) is a common paediatric vasculites potencially involving the heart, most significantly leading to the development of coronary artery aneurysms (CAA). The early institution of intravenous immunoglobulin (IVIG) is known to reduce this risk, however, response to this terapy is not uniform. Resistance to IVIG therapy can be predicted based on a scoring system, permitting the anticipation of adjuvant therapies. However this prediction model has not been validated outside the Japanese population. The aims of this study was to characterize the cardiac complications of KD and to identify predictors of IVIG resistance in our population. Retrospective analysis of KD cases diagnosed between January 2006 and July 2018 followed up in a Paediatric Centre.
Forty eight cases cases of KD were included, 67% were male, with a median age of 36 months (IQA 17-89). Twenty two (46%) had echocardiographic changes. Coronary involvement was found in 12 (25%): 5 CAA and 7 coronary ectasies. In the acute phase, 10 had pericardial effusion, 3 mild mitral valve regurgitation and 3 ventricular dysfunction, one with cardiogenic shock. One presented with variable atrioventricular block. After the acute phase, one patient maintained left ventricular (LV) dilation and conduction's system involvement and one had LV hypertrophy. Among the variables tested as predictors of IVIG resistance, C-reactive protein (CRP) showed an AUC ROC of 0.789 (IC95% 0.632; 0.947) and the erythrocite sedimentation rate (ESR) an AUC ROC of 0.781 (95% CI 0.585, 0.977). Cutoff points for CRP were 15.1 mg/dL with a sensitivity (Sn) of 0.778 and specificity (Sp) of 0.789 (OR = 13.125 IC 95% 2.271; 75.858), and for ESR 90.5 mm/h, with Sn 0.667 and Sp 0.857 (OR = 12.000 IC 95% [1.718; 83.803]). The logistic model with both variables presented was $p = 0.042$ and AUC ROC of 0.790 (95% CI [0.589; 0.992]). At the optimal cut-off point the Sn was 0.833 and Sp was 0.771.
About half of the patients had some form of cardiac involvment (coronary in 25% of the cases). Our results suggest that CRP and ESR values could be used as good predictors for IVIG resistance.

P-42
Role of congenital immunity genes in children with congenital heart disease undergoing cardiac surgery

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Introduction: Congenital heart disease (CHD) is one of the most common fetal malformations resulting in high postnatal disability and mortality. Heart surgery is one of the main methods of treating CHD. About 30% of patients develop postoperative complications. The innate immune response plays a special role in the development of complications. Immune response genes are associated with ontogenesis and their mutational variability may be accompanied by developmental defects. TREM-1, a key receptor for the innate immune response, is a central player in inflammatory response.
Purpose: To determine allele frequencies of the TREM-1 gene in children with congenital heart disease.
Materials and Methods: The study design was approved by the Local Ethics Committee. 154 children (81 girls and 73 boys) with congenital heart disease were included in the study. Patients were included in the study group after the diagnosis was confirmed according to the standard examination protocol (questioning, ECG, ECHO-CG). All the patients were assigned to two groups: children with ductus-dependent CHD and ductus-independent CHD. The mean age of patients was 6 years (from 5 to 8 years). Peripheral blood was collected from the cubital vein into tubes containing K2EDTA. Genomic DNA was extracted using phenol-chloroform. The TREM-1 8 loci (rs1817537, rs3804277, rs6910730, rs7768162, rs2234246, rs4711668, rs9471535, rs2234237) were genotyped with RT-PCR using Taqman probes. Statistical analysis was performed using the software packages SNPstats, GraphPad Prism.
Results and Conclusions: The distribution of allele and genotype frequencies conformed with Hardy-Weineberg equilibrium expectation. The T allele of the rs2234246 polymorphism was more frequently found in the group of patients with ductus-dependent CHD compared with the ductus-independent CHD (14.7% vs. 34.6%, $p = 0.022$, dominant inheritance pattern). The T allele of the rs4711668 was more frequently detected in the ductus-independent CHD (76% vs. 92% $p = 0.02$, dominant inheritance pattern). Importantly, patients with ductus-dependent CHD commonly had complicated postoperative period. TREM-1 is an inducer of the systemic inflammatory response; therefore, it seems promising to type TREM-1 for predicting postoperative complications after heart surgery for CHD.

P-43
Cardiac stress biomarkers in neonates: role of the delivery mode

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Introduction: Perinatal asphyxia is a common problem and is a significant cause of neonatal mortality and neurological as well as cardiological morbidity. Labour dystocia is the most common cause of perinatal asphyxia.
Several cardiac biomarkers such as cardiac troponin (cTn), CTnI, high sensitive-C-Reactive Protein (hs-CRP), copeptin and N-Terminal-pro-Brain Natriuretic Peptide (NT-pro-BNP) have been tested as possible indicators of perinatal asphyxia and neonatal morbidity. However, reference values in neonates are lacking.
The objective of our study was therefore to determine the reference value of of CTnT, CTnI, hs-CRP, copeptin, and NT-proBNP in healthy full term newborns and to test the hypothesis

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

that the type of delivery would influence cord blood concentrations of each biomarker.
Patients and methods: Cord blood samples were collected from 201 neonates delivered by uncomplicated vaginal route (n = 157), instrumental vaginal route (n = 18), scheduled caesarean section (n = 12), and urgent caesarean section (n = 14). Data on gestation, birth weight, sex, Apgar scores and respiratory status were recorded.
Results: Using the 99th percentile, the upper reference limit in healthy newborns was 49,59 ng/l for CTnT, 11,28 μg/l for CTnI, 1624,84 ng/l for NT-proBNP, 1301,40 pmol/l for Copeptin and 39,9 mg/l for hs-CRP. Neonates born after complicated delivery had significantly higher values of CTnT (P=0.001), CTnI (P=0.000) and Copeptin (P=0,008) than those born after normal delivery. hsCRP and NT-pro-BNP were not different between groups.
Neonates born by scheduled caesarian section showed significantly lower Copeptin concentrations than the other subgroups (P=0.01, respectively).
In a multiple regression model where Troponin T concentration was the dependent variable, the delivery mode was the statistically significant independent variable.
Conclusion: In this study, we established reference values of cord blood concentrations of cardiac stress biomarkers in healthy newborns. We showed that cardiac-related birth stress is dependent on delivery mode.

P-44

The congenital glycosylation disorder PGM1-defiency can give rise to severe cardiomyopathy and unexpected sudden cardiac death in childhood

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Introduction: Sudden cardiac death (SCD) in the young is rare, and should always lead to suspicion of a genetic cardiac disorder. We describe a family, in which the proband was a girl at thirteen years of age deceased by sudden cardiac death in the playground. The girl, the index-patient, had short stature, cleft palate but no previous cardiac symptoms. We found an uncommon cause of restrictive cardiomyopathy, due to a congenital disorder of glycosylation (CDG), previously described to cause a variable range of usually mild symptoms, and not previously found to cause SCD as the first symptom of the condition.
Methods: The index patient underwent postmortem genetic testing for genes known to cause SCD without a known causative agent. There was two siblings of similar phenotype as the deceased sister, why they underwent whole-exome genetic sequencing. All first-degree relatives underwent clinical examination including biochemistry panel, cardiac ultrasound, Holter-ECG and exercise stress test.
Results: A genetic variant in the gene for phosphoglucomutase 1 (PGM1) was identified in these two boys and the deceased sister, all were found to be homozygous for the genetic variant NM_002633.2:c.689 G>A in PGM1. This variant has been linked to a congenital disorder of glycosylation (CDG-PGM1), explaining the clinical picture of short stature, cleft palate, liver engagement and cardiomyopathy. During follow-up one of the brothers died unexpectedly after physical exertion during daily life at the age of twelve years. The other brother fainted during similar circumstances at the age of thirteen years. Both parents and three other siblings were found to be heterozygous gene carriers without risk for the disease.
Conclusion: Our findings suggest that there is a need of multidisciplinary discussion and the value of genetic testing after unexpected cardiac death in the young. In the global migration period we have to be more open-minded even for uncommon diseases as well as increased amount of recessive inherited disorders. Our findings suggest that the autosomal recessive CDG-PGM1 is highly associated with life-threatening cardiomyopathy with arrhythmia or sudden cardiac death as the first symptom presenting from childhood and adolescence.

P-45

Pathology and genetics of alveolar capillary dysplasia with misalignment of the pulmonary veins and its association with congenital heart defects

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Objective: Alveolar capillary dysplasia with misalignment of the pulmonary veins (ACD/MPV) is a rare and lethal congenital lung disease characterized by severe pulmonary arterial hypertension (PAH) and refractory hypoxemia. The mortality is nearly 100%, irrespective of the co-morbidities. The objective is to describe the pathology, genetics and clinical course of two patients with ACD/MPV and its potential impact on clinical decision-making.
Methods/Results: We retrospectively reviewed the records of all patients with ACD/MPV in our institution from 2015 to 2018 and the current literature. In this time period, we treated two patients. In both cases, a coarctation was seen. In the second case, an unbalanced partial atrioventricular septal defect (pAVSD) was detected as well. In both cases, first symptoms appeared during the first 24 hours of life. Main symptoms were PAH and refractory hypoxemia. X-Ray and cardiac catheterization were not conspicuous for a pulmonary vascular pathology. In both cases, aggressive management of PAH was not sufficient to ensure oxygenation. Thus, extracorporeal membrane oxygenation (ECMO) became necessary. However, patients could not be weaned from ECMO and died after ECMO therapy of 11 and 18 days due to complications, respectively.
We performed autopsy and array analysis for genetic testing. Histopathology revealed classical signs of ACD/MPV with reduced numbers of alveolar capillaries located away from the alveolar epithelium and malposition of pulmonary veins adjacent to pulmonary arteries. Array analysis showed de novo deletions in the chromosomal region 16q24.1q24.2. Both deletions included

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

the FOX Gene Cluster FOXF1, FOXC2 and FOXL1. Loss of function mutations of FOXF1 as well as deletions of the FOX gene cluster have been reported as disease associated.
Conclusions: Initially, ACD/MPV can be easily confused with persistent pulmonary hypertension of the newborn. This can prolong time to confirm diagnosis. Thus, ACD/MPV should be suspected in neonates with congenital heart disease and unexpectedly elevated pulmonary vascular resistances, especially in cases of obstructive left heart disease or AVSDs. It is important to increase the awareness of physicians. Further, it is crucial to perform lung biopsy and genetic testing at an early stage and to counsel the parents regarding future pregnancies.

P-46
Hydroxylapatite deposition induces graft alterations in CHD patients

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Objectives: Calcification-dependent graft failure often causes repeated operations in patients with congenital heart disease. However, the underlying patho-mechanisms are still poorly understood. Therefore, we analyzed the chemical composition of depositions in explanted tissue samples and compared them with serum levels of different ions. We aimed at developing prognostic parameters predicting graft longevity.
Methods: Different grafts explanted during pediatric cardiac redo procedures 10.69±8.65 years after initial implantation were examined. These included Contegra® grafts (n=3), homografts (n=2), one pericardial valve and one GORE-TEX® graft. Perioperative patient data and serum levels of potassium, sodium, calcium and phosphate were collected.
Micro X-Ray Fluorescence (μXRF) was used to detect the spatial distribution of different elements. Thin sections (10 μm) were prepared on silica slides and documented by light microscopy. An instrument equipped with a Rh X-ray tube (50kV, 30W) was operated with a spatial resolution of 25 μm. Five to ten cycles with a measuring time of 10 ms per point were performed and the spectra as well as spatial resolved images were created with the corresponding software.
For X-ray powder diffraction (Stoe StadiP, CuKα1 radiation, 60h counting time), highly crystallized areas were separated, washed with doubly-destilled water and dried in a vacuum desiccator. The dried samples were finely ground and filled into 0.3 mm glass capillaries.
Results: Severe macroscopic calcification was evident in all samples. Calcium and phosphorus were the main components of appositions encountered. Although the term calcification usually refers to calcium carbonate apposition, we could identify calcium phosphates instead.
The X-ray powder patterns revealed hydroxylapatite (Ca5(PO4)3OH), which is well-known as major component in dentin, tooth enamel and bone material. Calcium phosphates like alpha- and beta-Ca3(PO4)2 could be excluded.
Correlations of graft degeneration and clinical data, especially serum calcium levels, could not be proven at the time of reoperation.
Conclusion: Graft material is prone to hydroxylapatite deposition. Chemically, a similarity to bone matter and its degree of hardness could be shown. Whether medical treatment with chelating agents can improve graft longevity remains a question to answer. In this preliminary study, serum levels of ionized calcium did not correlate with the timing of graft failure.

P-47
Identification of intronic DNA sequences regulating titin expression in a dilated cardiomyopathy human induced pluripotent stem cell model

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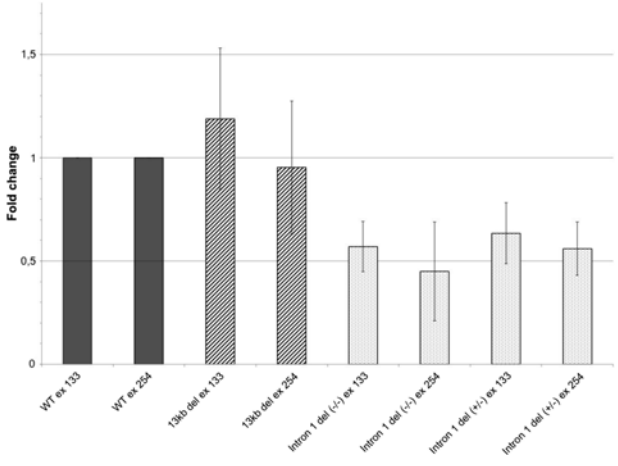
Introduction: Mutations that truncate the sarcomere protein titin (*TTNtvs*) are known to be a major cause of dilated cardiomyopathy (*DCM*). The aim of this study was to detect regulatory DNA regions that influence *TTN* gene expression. Manipulating those regions by microRNAs or small molecules could increase intact titin in cardiomyocytes and could provide new treatment options for *DCM*.
Methods: Deletions were implemented in human induced pluripotent stem cells (hiPSC) by transfecting with CRISPR/Cas9 guide RNA to target two putative regulatory regions, previously identified on bioinformatics analysis. Cloned cells were expanded and differentiated to iPS cardiomyocytes. After verification of *TTN* deletions by next generation sequencing, real-time quantitative PCR (qPCR) was applied to assess *TTN* RNA expression in heterozygous and homozygous mutated in comparison to WT clones.
Results: One deletion spanned 13,000 basepairs (13kb) upstream of *TTN* exon 1 excluding the TATA box and included a previously known SNP as well as a superenhancer region. Another separate deletion targeted a highly conserved region within Intron 1 of *TTN*. qPCR performed for two separate DNA sequences in exon 133 and exon 254 showed no significant difference in relative *TTN* expression compared to WT clones (see grey bars in Figure) in iPS derived cardiomyocyte clones carrying the 13kb deletion upstream of *TTN* (see striped bars in Figure). In contrast, homozygous and heterozygous deletions in Intron 1 significantly decreased *TTN* gene expression (see dotted bars in Figure; p-values: Intron 1 del (-/-) ex 133=0,02, Intron 1 del (-/-) ex 254=0,05, Intron 1 del (+/-) ex 133=0,04, Intron 1 del (+/-) ex 254=0,02) in iPS derived cardiomyocytes.



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusions: *TTN* Intron 1 plays an important role in *TTN* gene expression regulation and might be a relevant treatment target in *DCM* caused by titin-truncating mutations (*TTNtvs*).

P-48

Lessons from exome sequencing in prenatally diagnosed heart defects – a basis for prenatal testing

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Objectives: We aimed to illuminate the clinical utility of exome sequencing (ES) in cases with prenatally diagnosed congenital heart defects (CHDs).
Methods: In the present study, we retrospectively analysed the diagnostic yield as well as the percentage of non-conclusive findings and incidental findings in 30 cases with prenatally diagnosed CHDs in which we performed ES. In most cases, ES was done as parent-child trios.
Results: A definite genetic diagnosis was established in 20% (6/30) of cases. Non-conclusive results were found in 13.3% (4/30) of cases. Incidental findings were reported in 10% (3/30) of cases. There was a phenotypic discrepancy between the reported prenatal and postnatal extracardiac findings in 40% (8/20) of individuals. However, none of these additional, postnatal findings altered the genetic diagnosis.
Conclusion: Our study shows that ES in prenatally diagnosed CHDs results in a comparably high diagnostic yield. There was a significant proportion of cases in which incidental findings and variants of unknown significance in known disease genes were found, as well as potentially pathogenic variants in novel disease genes. These kind of findings can bedevil genetic counselling and decision making for pregnancy termination. Our findings also contribute to the range of prenatal findings in genetic disorders, which are unknown in most cases.

P-49

Has the electrocardiogram still additional value in the diagnosis of patients with (suspected) Noonan syndrome?

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Introduction: Noonan syndrome (NS) is a genetic disorder characterized by short stature, typical facial features, a variable degree of developmental delay and cardiac abnormalities. The cardiac abnormalities can be electrocardiographic abnormalities end/or congenital heart defects (CHD), most often a valvular pulmonary stenosis (PS) or hypertrophic cardiomyopathy (HCM).
In this retrospective study we analysed whether the ECG has still an additional value in the diagnosis of patients with (suspected) NS.
Methods: In this single center retrospective study we identified all patients with genetically confirmed Noonan syndrome and an available ECG and echocardiogram before any cardiac surgical intervention.
Results: In 95 included patients, mutations were most frequently found in the genes PTPN11, SOS1 and KRAS. ECG abnormalities were found in 60 patients (63,2%). There were 44 (46.3%) patients with left axis deviation, 29 (30.5%) with small R waves left precordial and 5 (5.3%) with large S waves right precordial. An abnormal Q wave and wide QRS complex were both seen in 2 (2.1%) patients. Patients with a mutation in RAF1 gene had significantly more frequent large S waves right precordial ($p=0.002$), and patients with SOS1 gene mutation had more often wide QRS complexes ($p=0.006$). No significant differences in ECG abnormalities were seen between patients with involvement of PTPN11 gene and other genes. Patients with PS had more often a small R wave left precordial ($p=0.007$) than patients without a PS, and patients with a HCM had more often a left axis ($p=0.021$) than patients without a HCM. Of the 34 patients without a CHD 13 (38.2%) had ECG abnormalities. These abnormalities were only seen in patients with mutations in PTPN11, SOS1, KRAS, CBL and BRAF genes. There was a significant difference between the group with CHD and the group without CHD in frequency of left axis deviation ($p=0.018$) and small R waves left precordial ($p=0.019$), which disappeared when the patients with cardiac valve insufficiencies were excluded.
Conclusions: Specific ECG abnormalities can still contribute to the early diagnosis of patients with NS, but can mostly not predict the possible gene involved and/or the (precence of a) CHD.

P-50

3D analysis of heterotaxy in the mouse model: from the embryonic heart loop to complex congenital heart diseases

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Introduction: The left-right asymmetric morphogenesis of the heart is initiated during the embryonic process of heart looping. The molecular cascade at the origin of the left-right asymmetry in the embryo has been well established in the mouse model and is affected in human cases of heterotaxy. Laterality defects such as heterotaxy are challenging to phenotype, because of partial penetrance, the number of organs affected and the complexity of the anatomical variations in the heart. Thus, laterality defects are currently incompletely phenotyped in the mouse model, hindering understanding of the pathophysiological mechanisms. Recent advances in imaging and computational analyses have opened the possibility to quantify morphogenetic processes in 3D and combine different modalities to detect different aspects of the phenotype.
Methods: We present a complete framework for imaging in 3D the shape and cellular architecture of the heart during embryogenesis and at birth, but also its relation to other visceral organs and the main vessels. This is validated in a model of heterotaxy.
Results: Using High Resolution Episcopic Microscopy (HREM), we have established a procedure to quantify the 3D shape of the embryonic heart loop. By lightsheet microscopy, we have addressed asymmetries in cell behaviour during heart looping. To correlate variations in embryonic shapes with specific configurations of heterotaxy at birth, we have established a multimodality-imaging pipeline combining echography, micro-computed tomography (micro-CT) and HREM. In the same individual, the embryonic shape is analysed by echography, whereas the situs of visceral organs is imaged by micro-CT. Finally, 3D reconstructions of the heart by HREM can identify complex cardiac malformations at birth, using the anatomical criteria of the segmental approach.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusion: This strategy is applicable to analyse a variety of mouse mutants with laterality defects. From quantitative 3D analyses at multiple scales and multiple stages, it is expected to provide novel insight into the mechanism of congenital heart defects associated with heterotaxy.

P-51

Elamipretide treatment in an infant with Sengers syndrome

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Introduction: Sengers syndrome (SS) is a rare cause of early onset cardiac failure, congenital cataracts and hypotonia caused by homozygous disease-causing variants in the AGK gene. We recently diagnosed a boy that was homozygous for a founder mutation (p.Ile348AsnfsTer38) carried by approximately 1% of Icelanders. In the first few months he was noted to have severe hypertrophic cardiomyopathy, bilateral cataracts and significant hypotonia, but average published survival of children presenting at this age is 4.2 months. SS and Barth syndrome have overlapping phenotypes and both are thought to lead to depletion of mitochondrial cardiolipin. Elamipretide is a tetrapeptide which is known to target the inner mitochondrial membrane and thought to bind and reduce cardiolipin damage, and thus theoretically may be of use to both syndromes.
Methods: In collaboration with Stealth BioTherapeutics we have treated our patient with elamipretide for ~6 months (starting at 3 months) through a compassionate care protocol, in addition to established beta-blocker treatment. Here we summarize data from weekly clinical global impression evaluations and bi-weekly echocardiograms during the 6 months of treatment.
Results: Pharmacokinetic studies indicated that drug exposure was similar to other elamipretide trials. Our patient showed subjective improvement from the prior week evaluation in 13/21 visits (~62%) and only was felt to worsen on one occasion. His global score went from markedly ill to borderline ill during the treatment period. Specifically, our patient's cardiac condition improved in the first few weeks of treatment and has remained stable, with 53% increase in left ventricular internal dimension in end-diastole and stabilization of left ventricle septal and posterior wall thickness despite a 40% weight gain due to normal growth. There have been no obvious side effects that are attributable to elamipretide. The patient has demonstrated intermittent lactic acidosis and neutropenia which are thought to be related to his condition. The patient has now been discharged to home.
Conclusions: Our treatment of a single individual with SS suggests that elamipretide is well tolerated in SS and that treatment may have helped stabilized our patients cardiac function. Further studies are required to definitely define the role of elamipretide in SS.

P-52

A case of early diagnosis the mucopolysaccharidosis type I (Hurler's syndrome) in 2-month-old girl with acute myocarditis

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Mucopolysaccharidosis I-Hurler is the most severe form of a metabolic genetic disease caused by mutations of the IDUA gene on chromosome 4p16.3 encoding the lysosomal α L-iduronidase. Acute cardiomyopathy has been found in some infants younger than 1 y.o.
Objective: To present a rare clinical case.
Methods: Complex inpatient examination.
History of the disease: 2 months old patient was hospitalized to ED with severe cardiac and respiratory insufficiency symptoms, convulsions.
Laboratory findings: high cardiospecific enzymes level (CK 271 U/l, CK-MB 122,5 U/l, LDH 546 U/l, Troponin I 0,2013) and NT-proBNP (82681pg/ml).
ECG and Holter monitoring A multiply atrial tachycardia 195 b.p.m. was recorded.
Echocardiography: LVEDD 39-40 mm. LVEF 20%.
Anomaly of the coronary arteries was excluded by CTA.
Chest X-ray Cardiomegaly (CTR 63.2%).
The management included ALV, tube feeding, intravenous inotropes and loop diuretics, complex HF and antiarrhythmic therapy, intravenous immunoglobulinG.
Positive dynamics was noted in the form of arresting the multiorgan failure syndrome, reducing signs of heart failure.
According to laboratory data-lack of viremia, decrease cardiospecific enzymes' level, NT pro-BNP (9356 pg/ml), normalization level of troponin. By ECHO LVEF was increase to 33-34% and there was't heart chambers size dynamic.
Due to early age of debut, the polysystemic lesion, the absence of positive myocardial remodeling dynamics on the optimal drug therapy lysosomal diseases were included to differential diagnosis.
Results: Low α-L-iduronidase by tandem mass spectrometry were detected. We researched IDUA gene by non-radioactive direct sequencing and found homozygous mutation Gln63Term, that associated with Hurler's syndrome. External manifestations disease were absent until 5 months. There was no family history of cardiac or mitochondrial disease. Specific therapy (Laronidase) was prescribed. Allogenic bone marrow transplantation was performed at 8 month. After transplantation, normalization NT-proBNP (550 pg/ml), increase LVEF to 48%, a decrease LVEDD to 30 mm were registered.
Conclusions: This patient had early clinical debut of the Hurler's syndrome with severe heart failure. The highlights of the case is that we did't see typical clinical manifestations for mucopolysaccharidosis typeI. The key point for effective diagnostics was atypical clinical features of acute myocarditis and genetic testing. Dynamic monitoring of the patient was continued.

P-53

New pathologic modality for diagnosis of mitochondrial cardiomyopathy

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Introduction: Definite diagnosis of mitochondrial cardiomyopathy has been challenging, because it requires respiratory chain (RC) enzymatic assay, and genetic testing. Now, we devised the new diagnostic approach of pathologic study including electron microscopic and immunohistopathologic study.
Methods: Ten patients with HCM were studied. Using results for respiratory chain enzymatic assay and genetic testing the patients were divided into MCM(4 patients) and non-MCM(6 patients) groups. Electron microscopy and light microscopy was performed

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

using endomyocardial biopsy samples. In electron microscopy study, volume density within cardiac muscle cells (CM) of mitochondria [Vv(mit,CM)] were measured using a systematic random sampling design. Using stored Formalin-fixed paraffin-embedded tissue, quantitive immunohistopathologic study with monoclonal antibodies against mitochondrial electron transport chain complex (Complex I, II, and IV) was performed. Staining area of complex I and IV were devided by that of complex II, an internal control marker of mitochondrial R.C.Thus, Area[Complex I/II] and Area[Complex IV/II] were measured.
Results: In the MCM group Vv(mit,CM) was significantly higher than the non-MCM group (40.6±2.8%, 21.9±4.1%, $p<0.001$) and significantly reduced Area[Complex I/II] or Area[Complex IV/II] or both were found.
Discusion: Volume density of mitochondria using electron microscopy can differentiate between MCM and non-MCM cases. Furthermore, immunohistopathologic study also showed a new possibility for diagnosis of MCM, which maybe useful for first-line screening.
Conclusions: Immunohistopathologic study may be a good screening test for MCM.

P-54
Dysregulation of Notch signaling in cardiac cells of the patients with tetralogy of Fallot

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Introduction: Tetralogy of Fallot (TF) is the most common cyanotic congenital heart defect. The cellular and genetic mechanisms of this defect are obscure. Mutations in several genes important for heart development have been described including mutations in the genes related to Notch pathway. Now is clear that fine-tuned sequential activation of Notch genes is responsible for the proper heart chamber development. The aim of this study was to analyze the activity of Notch pathway in the cardiac mesenchymal cells derived from TF patients.
Methods: Cardiac mesenchymal cells were isolated from 42 patients with TF and from 14 patients with ventricular septal defects (VSD), which was used as a comparison group. Notch pathway was analyzed by estimating the expression of Notch genes and receptors as well as the main Notch target genes by qPCR. Differentiation and proliferation capability of the cells was also estimated. For in vitro Notch activation Notch-intracellular domain bearing lentiviruses were used. Notch activation in hypoxic and normoxic conditions of in vitro culture was analyzed.
Results: The cells derived from TF patients demonstrated different pattern of gene expression profiles of Notch related genes comparing to the cells from the patients with VSD. Notch activation level by classic targets HEY1/HES1 correlated with the level of NOTCH1 transcripts in the cells from the patients with tetralogy of Fallot. Both in vivo and in vitro high Notch activation level was associated with enhanced differentiation and proliferation capacity of the cells. Hypoxic condition caused a very moderate elevation in Notch signaling in the cells from the patients with Tetralogy of Fallot.
Conclusion: Our data suggest a contribution of dysregulated Notch to the pathogenesis of tetralogy of Fallot; elevated level of Notch signaling could contribute to the increased plasticity of cardiac mesenchymal cells derived from the patients with tetralogy of Fallot. Our data confirm that fine-tuned Notch signaling is one of the key factors responsible for the appropriate heart development.

P-55
Identification of genetic susceptibility loci for coarctation of the aorta in 205 Swedish families

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Introduction: Coarctation of the aorta (CoA) is a congenital narrowing of the aorta which may present as an isolated condition or in combination with other cardiac anomalies. Males are affected twice as often as females, and it most commonly occurs without associated syndromic features. Even though CoA is typically not inherited in a Mendelian fashion, familial segregation has been described, suggesting underlying genetic influence for development of CoA.
Methods: To identify genetic variants associating with non-syndromic CoA, we collected saliva samples and isolated DNA from 205 Swedish patients with the main diagnosis of an isolated pre- or juxtaductal CoA, requiring surgical correction. This population included patients with an associated bicuspid aortic valve, persistent left superior vena cava, secundum atrial septal defects, patent ductus arteriosus or small muscular ventricular defects. Exclusion criteria were other associated cardiac anomalies, known chromosomal abnormalities, syndromes or extracardiac abnormalities. DNA samples were also collected from the parents, generating in total 205 families whereof 194 complete trios. The samples were genotyped using the Infinium OmniExpressExome chip from Illumina. A total of 564 individuals passed quality control, including 122 affected boys, 69 affected girls and their parents. We then performed transmission disequilibrium testing (TDT) to identify associations between CoA and genetic variants.
Results: The TDT analysis revealed associations with single nucleotide polymorphisms in chromosome regions 4q32.1 (OR 2.15, p=1.08x10-5), 6q16.3 (OR 0.45, p=1.60x10-5) and 13q33.1 (OR 3.27, 2.14x10-5). In a TDT analysis stratified on sex, associations in the male group were found with variants located in 8q24.23 (OR 3.50, 1.04x10-5), 11q22.3 (OR 0.40, p=1.13x10-5) and 8p22 (OR 2.31, 1.95x10-5), while the top associations in the female group were with variants in 6p21.31 (OR 0.24, p=5.53x10-6) and 14q22.3 (OR 0.25, p=1.33x10-6).
Conclusions: Genome-wide association studies for CoA have not previously been undertaken with a similar number of trios, and this study reveals susceptibility loci that may be involved in the development of the condition. The results also suggest that the genetic associations with CoA differ between the sexes.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-56

Overexpression of muscarinic receptors in SIDS: genetic data

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Introduction: Sudden infant death syndrome (SIDS) is unpredictable and poorly understood. It remains the leading cause of death among infants aged between 1 month and 1 year.
Overexpression of muscarinic M_2 Receptors (M_2R) was observed in hearts of newborns deceased from SIDS, and more recently in the blood of infants who experienced idiopathic life apparent threatening event (iALTE), suggesting the involvement of the vagal system overactivity in these pathologies.
We explored a family in which 3 infants out of 6 deceased from SIDS between the ages of 6 weeks and 8 months. To note that the 3 other children and parents are doing well, but they also experienced 2 fetal deaths (figure 1).
Methods: We first analyzed the blood expression of M_2R as a biological parameter reflecting vagal overactivity in 7 members of this SIDS family: total RNA was blindly extracted from blood samples and reverse transcribed into cDNA. M_2R mRNA expression was measured by PCR.
In terms of genetics, we analyzed the data of the Next Generation Sequencing (NGS) of the exomes of the 8 members of this SIDS family.
Results: M_2R overexpression was found in the 3 infants deceased from SIDS, in 1 healthy child and in both parents, whereas the expression of M_2R was normal in the 2 other healthy children.
The analysis of exomes identified 3 genes that could be involved in SIDS: *CAV3, CACNA2D2* and *SCUBE2*. However, the scenario and the mode of transmission are difficult to define. The first hypothesis is towards a trigenism, with 2 genes inherited from father, and one gene from mother.
Conclusions: The exacerbated vagal response, biologically expressed by overexpression of muscarinic M_2R, could be a risk factor for SIDS.
For the first time in this family, the overexpression of M_2R in both parents suggests a genetic transmission of a biological phenotype of vagal overactivity in humans.
The development of the sequencing of human exomes will probably allow the identification of genetic risk factors involved in SIDS. A complementary study of the whole genome may link the M_2R overexpression and the genetics.

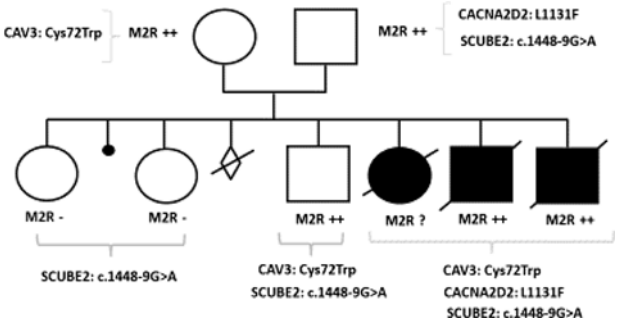

Figure 1.
Family tree of the « SIDS family » with expression of M_2R and data of NGS

P-57

Interleukin-1beta inhibition attenuates vasculitis in mouse model of Kawasaki disease

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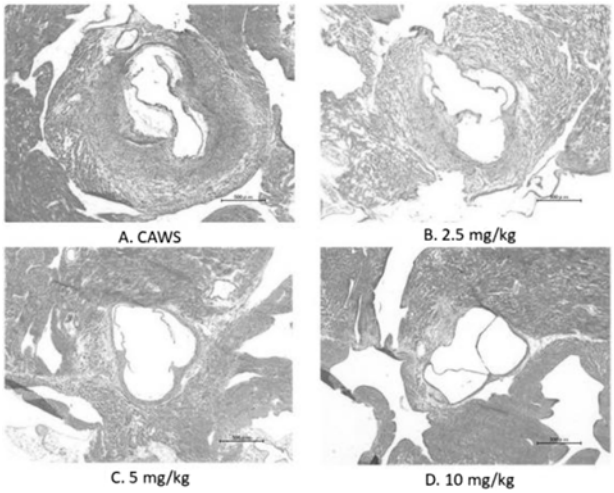
Introduction: Kawasaki disease (KD), a systemic vasculitis, is suspected to be related to abnormalities in innate immunity. Based on the importance of IL-1 signaling in innate immunity, we investigated the effects of an anti-IL-1β antibody using a Candida albicans water-soluble fraction (CAWS)-induced mouse model of KD.
Methods: CAWS (0.5 mg/mouse) was injected intraperitoneally into 5-week-old DBA/2 mice on five consecutive days. An anti-Murine IL-1β antibody (01BSUR) was administered at various doses (2.5, 5.0, and 10.0 mg/kg) and time points (2 days before, same day, and 2, 5, 7, and 14 days after CAWS administration). After 4 weeks, vasculitis in the aortic root was investigated histologically and serologically by cytokine profiling.
Results: Groups administered 01BSUR at all doses showed a significant reduction in the incidence of vasculitis. In addition, 01BSUR inhibited vasculitis until 7 days after CAWS administration. IL-1β, IL-6, and TNF-α levels were lower in 01BSUR-treated groups than in the group administered CAWS only. In an analysis of various time points, IL-6 levels were lower in all groups compared to the CAWS only group, but IL-1β, TNFα, and IL-10 levels were lower when 01BSUR was administered before CAWS. IL-10 levels tended to be higher when 01BSUR was administered after CAWS, suggesting that 01BSUR has additional effects beyond blocking IL-1β signaling.
Conclusions: The anti-IL-1β antibody significantly attenuated CAWS-induced vasculitis.



P-58

InsG791 mutation in cardiac myosin-binding protein C (MYBC3) gene: clinical characteristics in the Mennonite pediatric population, Manitoba, Canada

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Objective: Hypertrophic cardiomyopathy (HCM) is a common genetic and markedly heterogenous disease often associated with sudden cardiac arrest/death among young, otherwise healthy people.
It is caused by more than 1400 pathogenic mutations in one of at least 11 genes encoding sarcomere proteins [1].
A unique mutation, InsG791, in the gene for cardiac myosin-binding protein C (MYBC3), is found to be responsible for cases of familial HCM among the Mennonite community population in the Province of Manitoba. Therefore, we are interested in conducting a study to determine some of the clinical characteristics of this unique mutation identified in Mennonite children living in our province.
Methods-Results: Retrospective study includes 35 Mennonite children (M:16, F:19), aged between 0-18 years, carrying identical mutations (InsG791) in the gene MYBC3 and managed at the Variety Children's Heart Centre, Winnipeg, Manitoba. First echocardiogram was performed at median age 9.4 years (0.42-17.17). Five (5) were diagnosed with HCM in the form of septal hypertrophy on echocardiography. Incidental findings include Bicuspid Aortic Valve, small VSD and mild Pulmonic Valve stenosis in 3 non-HCM patients. All of the participants in this study remained asymptomatic throughout a median follow-up period of 7.5 years. No deaths were reported.
Conclusion: InsG791 mutation in the MYBC3 gene, identified in Manitoba's Mennonite pediatric population, appears to be associated with, later age of onset of the disease and better survival, in contrast to other FHC mutations. Half of the Canadian Mennonite population resides in the Province of Manitoba. As we have shown in a previous study, Mennonite pediatric population contributes approximately for 81% the of the total reported pediatric HCM cases in the Province of Manitoba [2]. Therefore, we believe that it could have implications on diagnostic considerations and screening strategies for the children with this unique mutation in our province.

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P-59
Pediatric cardiac transplantation after Fontan failure: A single-institution experience

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Introduction: Despite the excellent outcomes in the current era after the Fontan operation, it continues to have an inherent risk of failure. Cardiac transplantation could be an option for treating these patients; however, the indications for, timing of, and outcomes after, transplantation remain undefined. We reviewed our own institutional experience with transplantation for failed Fontan.
Method: The medical records of pediatric patients with heart transplantation in Asan Medical Center, since 1997 were reviewed retrospectively. Among the 72 patients who underwent cardiac transplantation (42 males and 30 females, median age at transplant is 12.6months), 7(9.7%) had previous history of Fontan operation (3 male, 4 female). Extracardiac conduit Fontan procedure was performed in all patients.
Result: For 7 eligible patients, the median age at Fontan procedure and heart transplantation is 3.8 years and 14.3 years, respectively. The mean interval from Fontan to transplant is 120.4 months. The most common indications for transplantation is ventricular dysfunction (6 patients, 85.7%), and 1 patient had uncontrolled protein-losing enteropathy (PLE). Only 1 patient shows high value of panel reactive antibody (PRA) before transplantation. When transplant was performed after desensitization therapy, no sign or symptom of acute rejection was seen (4 months of follow up). The mean duration of follow up is 41.1 months. One expired from acute rejection (42.4 months after transplantation), 1 had Cytomegalovirus infection and 2 had tacrolimus induced Diabetes Mellitus. No other significant complication associated with cardiac transplantation was showed.
Conclusion: Transplantation is an acceptable treatment for patients with a failed Fontan. As the numbers of patients with Fontan operations are increasing, the number of children, adolescents, and young adults requiring late rescue therapy with heart transplantation will increase. With the sparse availability of hearts for transplantation, careful allocation of recipient who underwent Fontan procedure will be need. And moreover, multi-centered large scale study for pre and post-transplant management appropriate for those patient should be performed.

P-60
Retrospective Analysis of Thrombosis and Bleeding Events in the Children with implanted Ventricular Assist Device

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Introduction and objectives: When the drug therapies do not work in the heart failure, mechanical support like ventricular assist devices are used. They improve quality of life, reduce the hospital stay and provide bridging heart transplantation safely. The aim of this study is to evaluate the bleeding/thrombosis complications in our left ventricle assist device implanted patients.
Methods: The left ventricle assist device implanted children who are followed up between June 2009-August 2018 were enrolled for the study. Patients were evaluated retrospectively using their demographic datas, preimplantation characteristics and especially bleeding/ thrombosis complications.
Results: Participants were twenty-six children who are aged under eighteen years; they are divided into two groups. One group has the patients to whom Berlin Heart Excor pulsatile assist device was implanted; the other group has the patients to whom HeartWare continuous assist device was implanted. 57.5% participants (n=15) were male, 42.3% were female (n=11). Median age was 185.0 months (min 28-max 295 months). All the subjects were diagnosed as dilated cardiomyopathy. 10 of them (38.5%) had underlying diseases which were glycogen storage disease type IV(n=1; 3.8%), Becker muscular dystrophy (n=2; 7.7%), non-compaction cardiyomyopathy (n=4; 15.4%), transposition of great arteries (n=1; 3.8%) and arrhythmogenic right ventricular dysplasia (n=2; 7.7%). 14/26 had thrombosis event at least once; whether critical or not (53.8%); 8/26 had bleeding event at least once; whether life-threatening or not (30.8%). 10 out of 26 never had an event. 32 important events recorded as 34.6% pump thrombosis, 30.8%

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

stroke, 11.5% intracranial bleeding, 3.1% MI and 6.2% intraventricular thrombosis. 16 events were not life threatening (33.3%). Statistically difference is not found between two device groups according to complications (p=0.199).
Conclusion: The results cannot be generalized in consequence of unequal dispersion of the groups. Nearly the same outcomes are obtained compared to other multi centered studies. To lessen the complications as thrombosis and bleeding, yet there is not a consensus used for the management of pediatric ventricular assist device implanted children. In order to balance status between bleeding and thrombosis the personal characteristics of patients should be well known, drug doses should be followed closely.

P-61
Therapeutic options and survival of patients with plastic bronchitis after single ventricle palliation – a systematic review

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Introduction: Plastic bronchitis (PB) is a rare complication in single ventricle (SV) patients of which the exact pathophysiology, survival and best treatment are still unclear. This study aims to systematically review the literature to give insight into characteristics, survival and management of SV patients with PB.
Methods: A systematic review was conducted, using the PUBMED database, to find articles, published up to August 2018, with SV patients and PB of which characteristics, treatment and/or outcome were well described per case.

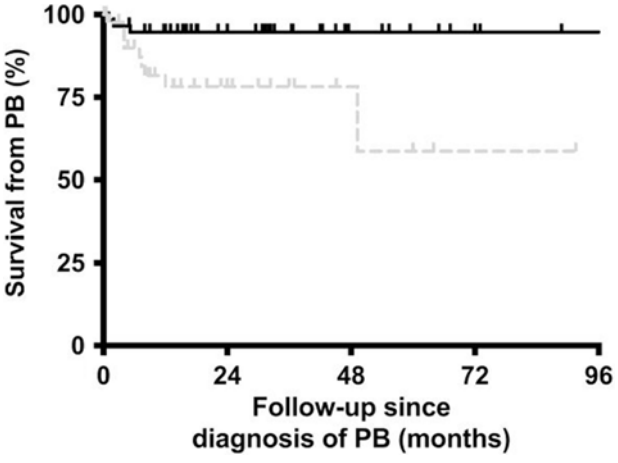

Figure.
Kaplan-Meier curve of survival of patients with plastic bronchitis (PB) based on period of diagnosis after Fontan palliation, diagnosis within 12 months (light grey dashed line) and after 12 months (dark grey solid line) after Fontan palliation.

Results: 576 articles were screened and 72 articles had sufficient data of 132 well described SV cases with PB. Most cases had a Fontan palliation (n=125) with a median age at diagnosis of PB of 60.0 months (IQR 41.0-85.3), median age at Fontan operation of 36.5 months (IQR 25.5-50.4), median interval between Fontan operation and diagnosis of PB of 18.0 months (IQR 5.0-36.6) and a median follow-up after diagnosis of PB of 18.0 months (IQR 6.5-36.8). Mortality was 15.2% (n=20) with a median period of 3.5 months after diagnosis of PB. Patients were treated with a combination of medical and interventional/surgical treatment (n=101), only medical (n=13) or only interventional/surgical treatment (n=11). Most reported drugs were fibrinolytics (n=75), most described intervention was bronchoscopic cast extraction (n=61) and most reported catheterization and/or surgical treatments were ligation/embolization of thoracic duct (n=33), relief of arterial, venous or intra-cardiac stenosis (n=26), creation, dilation or stenting of fenestration (n=24), and occasionally Fontan takedown or heart transplantation. Mortality was associated with diagnosis of PB within 12 months after Fontan palliation versus diagnosis after 12 months after Fontan palliation (five-years survival of 56.1% within 12 months vs 94.7% after 12 months, p=0.003; Figure) and a higher age at Fontan operation (47.4 months in the mortality group vs 36.0 months in the survival group, p=0.013).
Conclusions: Most cases are diagnosed with PB one year and a half after the Fontan palliation and around one-sixth of the cases die after a short period after diagnosis. A negative outcome is associated with diagnosis of PB within 12 months after Fontan palliation and a higher age at Fontan operation.

P-62
Experience with LENUS pro® pump implantation in two pediatric patients

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Introduction: Prostanoid treatment in patients with severe pulmonary arterial hypertension has been proven safe and effective. Subcutaneous administration of treprostinil associates side effects which limit their use and acceptance. A fully implantable pump for continuous application of intravenous treprostinil has been approved in Germany. We describe our experience with the implantation of this pump in two pediatric patients with severe idiopatic pulmonary hypertension.
Description of Cases: The LENUS pro® pump was implanted in two fifteen year-old patients with severe idiopatic pulmonary arterial hypertension. Both treated previously with tadalafil, ambrisentan and subcutaneous treprostinil. In both patients the indication for Lenus pro® pump implantation were local side effects such as pain and inflammation that were not well tolerated and decreased severely the quality of life of our patients. The pump was surgically implanted under general anesthesia without complications.

In the postoperative period one patient had a neumothorax and was hemodinamically instable requiring vasoactive drugs. Treprostinil was administered at 40 ng/Kg/min through the pump plus an additional subcutaneous treatment that was gradually increased up to a total dose of 85 ng/Kg/min, due to cardiac deterioration. This patient had been previously

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

refused for lung transplant. The patient presented repeated pulmonary hypertension crisis deceasing at day 9 after the pump implantation.

The second patient was discharged 4 days after pump implantation with treprostinil at 60 ng/kg/min and no subcutaneous infusion. The doses was gradually increased up to 92 ng/kg/min, well tolerated and with no complications. This patient has been followed for 18 months in which he refers an improvement of his quality of life.

Comments: Implantable pumps for parenteral prostanoid administration in pediatric patients are an alternative to external pumps, especially when familiar, psychological or psychomotor issues hinder the use of external pumps. However, the risk associated of general anaesthesia should be considered previously.

P-63

Alveolar capillary dysplasia with misalignment of the pulmonary veins: two cases associated with congenital heart disease

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Introduction: alveolar capillary dysplasia with misalignment of the pulmonary veins (ACD/MPV) is a rare and lethal cause of pulmonary hypertension (PH) in newborns, frequently associated with cardiovascular and other systems anomalies. We present two cases of neonates with congenital heart diseases and refractory PH with post-mortem diagnosis of ACD/MPV.

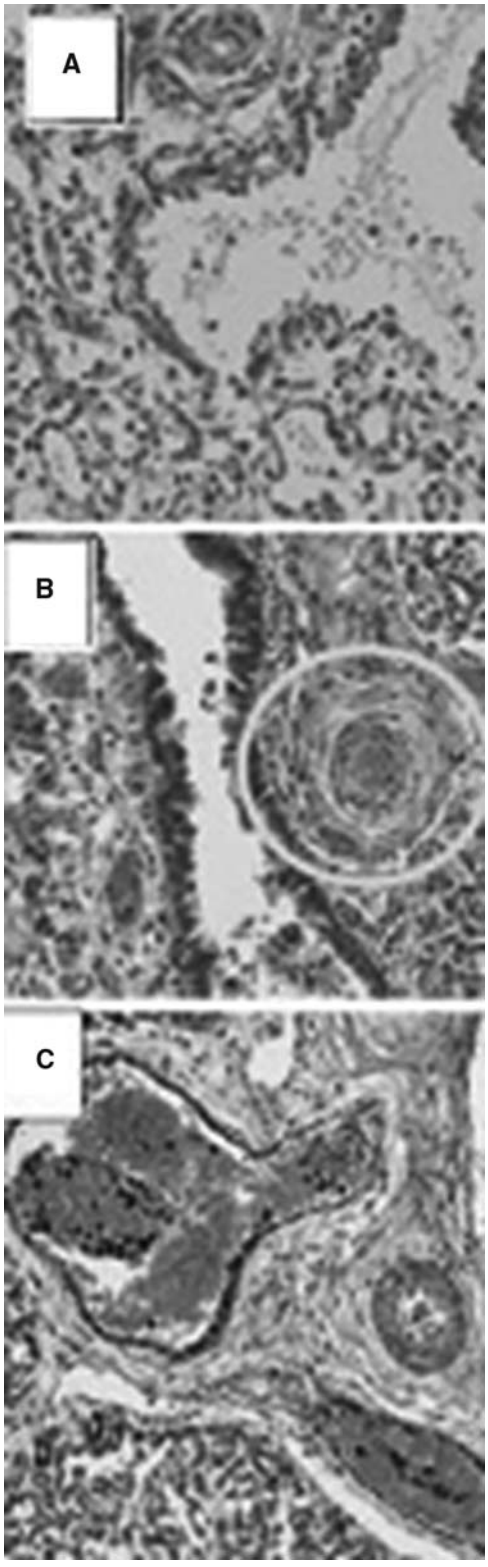
Methods: retrospective review of patient records regarding clinical findings, image and genetic tests, and autopsy data.

Results: the first patient was a full-term male who developed respiratory distress requiring mechanical ventilation within his first hours of life. Extubated on day three, he began suffering hypoxic episodes. Echocardiogram and cardiac CT showed coarctation of the aorta with arch hypoplasia, signs of pulmonary hypertension, large patent ductus arteriosus and a levoatriocardinal vein. Non-invasive respiratory support and prostaglandin infusion were started. Aortic arch repair and levoatriocardinal vein ligation were performed on day 15. After surgery, he suffered severe pulmonary hypertensive crises refractory to maximal ventilatory parameters and pulmonary vasodilator treatment. Cardiac catheterization demonstrated suprasystemic pulmonary artery pressure with normal PWP. ACD/MPV was suspected, but the patient developed multiorgan failure and died on day 22 before performing pulmonary biopsy.

The second case was a full-term male with prenatal diagnosis of complete AVSD initially discharged without treatment. He was admitted again on day 29 due to hypoxic episodes, starting oxygen therapy and anticongestive treatment. Echocardiograms showed unbalanced AVSD and signs of severe PH. *His clinical status progressively worsened*, requiring mechanical ventilation on day 35. A cardiac CT ruled out pulmonary vein stenosis, and cardiac catheterization demonstrated suprasystemic pulmonary artery pressure with normal PWP. Pulmonary vasodilators were initiated, with initial but unsustained response. As ACD/MPV was suspected, genetic test was performed and pulmonary biopsy was programmed, but he suffered a left ACM stroke on day 59 and limitation of the therapeutic effort was performed.

Both necropsies and genetic tests were compatible with ACD/MPV ***(Table 1).***

Conclusions: ACD/MVP must be suspected in neonatal cases of refractory pulmonary hypertension without typical risk factors. The association with congenital heart disease must not mislead the clinical suspicion. Lung biopsy should be performed to consider limiting invasive procedures and provide palliative care.



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

	Case 1	Case 2
Pulmonary findings	Underdeveloped alveoli and diffuse expansion of the interstitium (a); capillaries away from alveolar basement membranes. Thick and tortuous arterioles, with a marked narrowing of the lumen by intimal hyperplasia and fibrinoid necrosis (b, circle). Large and dilated veins, abnormally located beside the central bronchiolar-vascular bundles (c). ***Figure 1 (Patient 1)***	
Associated anomalies	Aortic coarctation with arch hipoplasia and bicuspid aortic valve. Bilobed right lung, symmetric liver and intestinal malrotation.	Complete unbalanced AVSD. Intestinal malrotation.
Genetic findings	De novo hemizygous deletion of chromosome 16q24.1-q24, containing FOXF1, FOXC2, FOXL1 and JPH3 genes.	De novo heterozygous variant in FOXF1: c.257G>C (p.Arg86Pro).

P-64
Switch from bosentan to macitentan in children and adolescents with pulmonary arterial hypertension during a hospitalization period

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Introduction: Macitentan is the most effective of the known endothelin receptor antagonists approved for the treatment of pulmonary arterial hypertension (PAH) in adult patients. Radomized controlled studies have shown that matsitentan improved functional class, long-term outcomes, reduced disease progression and mortality, as compared to previous therapy. In children, the application experience is limited to a few small studies.
Materials and methods: during the period from June 2018 to December 2018, 8 patients, including 3 boys and 5 girls aged 13.4+ 7 years, including 2 – LAG-CHD, 6 – ilag, who received therapy with bosentan and sildenafil for 2 to 7 years, were switched frombosentan+sildenafil to the macitentan and sildenafil therapy. The reason for the change of therapy was the worsening of the patient's condition in the presence of the maximum therapeutic doses of sildenafil and bosentan. All but one patient received 10 mg of matitanana a day.. One girl of 9 years with idiopathic PAH received 5 mg of matitanan per day. Duration of treatment in cardiology department was from 7 to 10 days.
All patients underwent standard laboratory tests, liver ferments analysis, ECG, ECHOCARDIOGRAPHY, 6-minute walk test, cardiopulmonary test.
Results: the functional class decreased from 3.1+0.64 to 2.6+- 0.74 (p=0.17), the distance at 6 minutes walk test increased from 390+99 to 494+111 m (p=0.067), the excursion of the fibrous ring of the tricupid valve increased from 17.5+4.8 to 20.5+4.9 mm per second (p=0.23)
Conclusions: the change of therapy in children with primary pulmonary hypertension and pulmonary hypertension associated with congenital heart disease was performed. Combination therapy with sildenafil + bosentan was switched to sildenafil+macitentan. There was a decrease in the liver enzymes level. This switching is safe and effective. This is confirmed by short-term observation in a cardiological department. Such a switchover can improve the condition of children with PAH without joining the therapy of the third specific drug.

P-65
Favorable outcome after heart transplantation in children: 18 years' experience of the Dutch national transplantation program

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Objectives: Heart transplantation (HTx) in children is concentrated in one center in the Netherlands. The objective of the study was to evaluate the outcome of a national pediatric heart transplantation program.
Methods: A retrospective, single-center, descriptive study of all children listed for heart transplantation in the Netherlands between 1999 and 2017.
Results: In total, 69 children were listed for HTx at a median age of 10.3 years (IQR 2.5-13.1). The underlying diagnoses included dilated cardiomyopathy (n=43, 62%), restrictive cardiomyopathy (n=8, 12%) and congenital heart disease (n=3, 4%), and 15 (22%) had another etiology. At HTx listing, (n=52, 75%) patients were hospitalized. Forty-seven (68%) children were successfully transplanted at a mean age of 10.5 ± 4.8 years. Outcome after HTx was favourable, 5-years survival was 95% (n=43) Three children died after a mean of 4.0 ± 3.5y ears after HTx, one in the 1st year from infectious complications after treatment for severe cellular rejections, two at adolescent age due to graft failure after rejections related to non-adherence. One patient underwent successful retransplantation. Twenty-two (32%) children on the waiting list were not transplanted: 16 (25%) died, 8 of whom on mechanical circulatory support (MCS), which was available in our program as of 2007. One patient was delisted with improved cardiac function after 9 months on MCS, and 4 patients were still listed at the end of the study. Overall, the functional outcome after HTx was favorable: all children returned to school, and participated in age-appropriate physical activities. In the majority of the recipients, graft function has been good at follow-up. The most important transplantation-related complications were post-transplant lymphoproliferative disease (n=8, 13%), cardiac allograft vasculopathy (n=3, 6%) and infections: Epstein-Barr virus infection (n=15, 32%) and Cytomegalovirus infection (n=10, 15%). No hemodialysis or kidney transplantation was needed.
Conclusion: A national approach allowed an adequate base for a pediatric heart failure - and transplantation program. We report favorable outcomes, in line with international standards. Heart transplantation is a viable option for a selected group of children with end-stage heart failure and without alternative treatment options. Currently, insufficient donor availability remains the critical limitation for transplantation.

P-66
Treatment of bronchopulmonary dysplasia-associated pulmonary hypertension with pulmonary vasodilators – single center experience

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Introduction: Patients with bronchopulmonary dysplasia-associated pulmonary hypertension (BPD-PH) have significantly higher mortality (up to 50%) and morbidity compared to patients with bronchopulmonary dysplasia (BPD).
Objective: The aim of this study was to present single center experience and outcome of the treatment with pulmonary vasodilators patients with severe BPD-PH

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Methods: BPD-PH risk factors (according to Nagiub et al), WHO-FC, NTproBNP, echo estimated RV systolic pressure before treatment and during follow-up, were chosen for retrospective analysis.
Material: Between 2009-2018 12 pts with severe BPD-PH diagnosed at age 9,4 mths (3-18,5) were accepted for treatment. Ten pts (83%) had severe BPD with oxygen blood saturation 70-90%HbO2. BPD-PH risk factors (2-7) were confirmed in all. Severe PH (RV systolic pressure >2/3 of systemic) was diagnosed in cardiac catheterisation in 8 pts and by echo in 4 pts. Seven pts were in III-IV WHO-FC and 5 pts in II WHO-FC. Median NTproBNP was 3000pg/ml (171-35000).
Congenital shunt coexisted in 6 pts: small PDA -1, ASD -2, VSD -2. Neonatal PDA closure was performed in 3 pts. 3 pts were diagnosed after shunt closure (2 ASD, 1 VSD). Pulmonary vein stenosis was excluded in all pts.
Treatment included oxygen therapy (11pts), sildenafil (11pts) and sildenafil + bosentan (1pts) with no side effects.
Results: Improvement in WHO-FC, NTproBNP after 3 mths of therapy was observed in 9 pts (75%). No changes in oxygen blood saturation were found.
4 deaths (33%) occurred 1,8-8,1 mths after beginning of therapy: 1 related to sepsis in patient without shunt and 3 in patients with VSD (1 after PAB, 2 after surgical VSD closure).
Pulmonary vasodilators were discontinued in 6 pts (50%) under echo/cardiac catheterization control after 10,3-40,7mths. 2 ASD were closed >1 year after treatment discontinuation. Two patients still require PH treatment.
Conclusions: BPD-PH treatment with pulmonary vasodilators is well tolerated, led to clinical improvement in majority of patients and normalisation of pulmonary hypertension parameters in half of survivals.
Decision on VSD surgery in patients with BPD-HP must take into account the high risk of postsurgical death.
Further studies with larger population are needed to establish outcome in patients with BPD-PH.

P-67
Prenatal echocardiography in a "typical" Tetralogy of Fallot

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Introduction: The underlying mechanism in the embryology of tetralogy of Fallot (TOF) is assumed to be the disproportional division of the conotruncus during early development of the heart. Unlike the postnatal definition, prenatally usually no pulmonary stenosis is found, and the diagnosis is described only by a ventricular septal defect with an overriding aorta. The aim of the study was to quantify the aorta and pulmonary artery prenatally in a "typical" TOF.
Patients and methods: Retrospective study of prenatally diagnosed TOF was performed. Included were only patients with normal-sized ventricles and unobstructed antegrade flow through the pulmonary valve (PV) and ductus arteriosus (DA); excluded were patients with atretic/dysplastic/absent PV or retrograde DA flow. Analyzed were 24 patients in 41 serial examinations; and data were compared with gestational week (GW) matched healthy controls (NORMAL). Analyzed were: aortic (AoV) and PV annulus Z-score according to GW, PV/AoV ratio, aortic arch (AA), DA, DA/AA ratio, right/left ventricular ratio (RV/LV), right/left ventricular wall ratio (RVW/LVW). Fetal echocardiography was performed between 20^{th}-40^{th} GW, with 6-12 week interval (median 9) between exam 1 and exam 2.
Results: Smaller PV (Z-score<2) in 85.4%; and bigger AoV (Z-score>2) in 53.6% of TOF patients were found. Comparing TOF/NORMAL significant differences were found (median) in: AoV Z-score 2.19/0.61 (P<0.0001); PV Z-score -2.98/-0.3 (P<0.0001); PV/AoV ratio 0.67/1.12 (P<0.0001) with cut-off point 0.9; DA/AA ratio 0.71/0.96 (P<0.0001) with cut-off point 0.85. There was no significant difference in RV/LV or RVW/LVW ratio. No significant differences during serial follow-up in any of the measured parameters were detected.
Conclusions: Our study confirmed in TOF patients already prenatally a disproportional size of the great arteries, with mildly dilated aorta and a smaller pulmonary artery, and this despite preserved and unobstructed antegrade pulmonary flow. On the contrary to other studies, we did not find any progression of this unfavorable setting during serial follow-up. In prenatal TOF establishing the patient´s own pulmonary artery / aorta ratio enables quick and easy evaluation of the pulmonary artery regardless patient´s size or gestational week. This may be of important prognostic value for the postnatal clinical course and further management.

P-68
Comparison Of Antenatally And Postnatally Diagnosed Atrioventricular Septal Defect (AVSD) And Its Contemporary Outcomes In South Wales

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Objective: To compare clinical associations and outcomes of fetuses and infants diagnosed with atrioventricular septal defect (AVSD) in South Wales.
Methods: Retrospective study of all antenatally and postnatally diagnosed AVSD in South Wales, between 2001 and 2018.
Results: 287 cases of AVSD were identified: overall 69.33% (n=199) were diagnosed antenatally (AN) and 30.66% (n=88) postnatally (PN). The antenatal pick-up rate increased over the years from 25% to over 60%. Karyotyping was available in 183 cases: 123 had trisomy-21 and 18 trisomy-18. Pregnancy was interrupted (TOP) in 90 cases (45.23%) and intrauterine death and stillbirth were noted in 20 cases.
Associated cardiac anomalies were found in 159/287 cases; 132 also had non-cardiac anomalies. Out of live births, 43/88 PN cases versus 65/89 AN cases had additional cardiac anomalies. Isomerism, unbalanced AVSD, pulmonary atresia and coarctation were significantly more common in the AN group, which translated into poorer outcomes with higher mortality rates (AN 35.96% versus PN 12.50%).
89 antenatally diagnosed patients were delivered alive: 65 had additional cardiac and 44 non-cardiac anomalies, compared to the TOP group (n=90), where 38 had additional cardiac and 40 had non-cardiac anomalies. Karyotyping was positive in 42 delivered patients versus 50 interrupted pregnancies. The most common cardiac anomaly was unbalanced AVSD for both groups (22.47%, n=20 versus 20.00%, n=18), followed by pulmonary atresia or stenosis (21.35%, n=19 versus 12.22%, n=11). Main non-cardiac anomalies were gastrointestinal (24.72%, n=22) for delivered patients and musculoskeletal (26.67%, n=24) for the TOP group.
101 underwent AVSD repair and 27 had complex surgical procedures for Fontan route. 29 required mitral valve repair (with four

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

requiring mitral valve repair redo and five mitral valve replacement). 6 required pacemaker implantation. Postop mortality rate was 19.28% (n=16) for the associated cardiac anomalies group and 5.17% (n=3) for the isolated group. Risk factors for increased mortality were antenatally diagnosis and additional serious cardiac anomalies.
Conclusions: Although the number of antenatally diagnosed AVSDs increased over the years, this trend had no favourable effect on the surgical outcomes. Karyotype, associated cardiac or non-cardiac anomalies had no influence on patients' decision for pregnancy interruption. The mortality rate for AVSD associated with complex cardiac lesions remains high.

P-69
Outcome of hydropic fetuses with supraventricular tachycardia need not be dismal
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Background: Prognosis of fetuses with hydrops and supraventricular tachycardia has been portrayed as poor in most published reports. This may lead to biased counselling, unnecessary Caesarean section, preterm delivery and even termination of pregnancy. However, when treated effectively even hydropic fetuses may have similar outcomes like non-hydropic ones.
Aims: To evaluate contemporary fetal and postnatal outcomes of hydropic fetuses with supraventricular tachycardia (SVT) when it is treated with effective antiarrhythmic medications and monitored systematically.
Methods: This is a retrospective review of single centre experience over a 15-year period. All fetuses received high dose flecainide and digoxin combination treatment. Tachycardia response rate, time to arrhythmia and hydrops resolution, fetal and postnatal morbidity and mortality rates were analysed.
Results: 24 cases had SVT, hydrops and signs of cardiac dysfunction. Two patients with medical termination and one case of intrauterine death were excluded from the study. Mechanism of SVT was atrioventricular re-entry tachycardia (AVRET including WPW and PJRT) in 14 and atrial flutter (AF) in seven cases. Median heart rate was 271 beats per minute (bpm) in AVRET and 400 bpm in AF.
Among the 21 fetuses treated, overall tachycardia response rate was 90% with restoration of sinus rhythm in 80% of the cases. Tachycardia responded to medication in 93% of AVRET with median conversion time of 1.5 days. In fetuses with AF tachycardia termination or rate control occurred in 71% of cases within a median of 1 day. Hydrops resolved in all SVT cases except in one fetus who had a along RP tachycardia and did not-respond to any other medications and even direct intrafetal treatment.
There was no maternal morbidity owing to medical treatment. Four fetuses went into spontaneous preterm birth and one fetus was delivered early due to worsening hydrops. No neurological morbidity was documented in surviving neonates. There was one postnatal death due to respiratory complications of prematurity in the non responsive long RP case.
Conclusions: High dose flecainide and digoxin combination offers a rapid and effective treatment strategy in fetuses with hydrops and supraventricular tachycardia. This report may aid more accurate counselling of hydropic fetuses.

P-70
Fetal Arteriovenous intrapulmonar Fistula Case Report
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Introduction: Fetal arteriovenous malformations are very rare, but they are diagnosed more and more.
The echocardiograms signs are dilatation of right pulmonary artery and right pulmonary vein, with cardiomegaly or not. Those with high flow can lead to heart failure and hydros.
Newborn patients can be asymptomatic at first, afterwards they develop cyanosis.
The diagnosis is confirmed postnatally, generally throught an Angio tomography, and the correct treatment is percutaneous closure (device)
Case Report: A 30 years old pregnant woman, fetal echocardiogram was performed at 27 weeks of gestational age, and dilatation of right pulmonary artery and right pulmonary vein were found.
Results: At birth a systolic murmur and slight cyanosis were present. An angioTac image confirmed the presence of an AV intrapulmonary RPA-AI Fistulae. At 18 days of life and with 3800 kg, the fistulae was succesfuly closed with an vascular plug device.
Conclusion: The fetal arteriovenous malformations are very rare but must be suspected in the presence of dilatated cardiac structures, even more if heart failure and hydros are present.

P-71
Shared Decision-Making Between Physicians and Parents of Children with Hypoplastic Left Heart Syndrome: A Qualitative Study of the Communication Challenges Associated with Therapeutic Evolution.
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Introduction: Children with hypoplastic left heart syndrome form a heterogenous group with significant risk of morbidity and mortality. This leads to uncertainty over the preferred course of care due to difficulty assessing the benefit of therapeutic intervention. Physicians are faced with engaging parents in a shared decision-making process that is both emotionally and cognitively complex. This study seeks to identify common features in the approach of paediatric cardiologists and trainees to communicating with parents in this context, and to ascertain their perception of post-graduate training for such communication.
Methods: This was a qualitative descriptive study. Data was collected using two focus groups of consultants and trainees practicing in paediatric cardiology. Transcriptions were coded using the NVivo Software programme. Thematic analysis was used for analysis and interpretation.
Results: Two separate focus groups were conducted with trainees and consultants, consisting of three participants each. Three central themes were identified; the delivery of medical information, building a relationship with parents and learning from experience. Consultants in our focus group placed more emphasis on social circumstances and developing relationship of trust with parents than was apparent in the trainee focus group. Both groups described limited formal communication training relevant to this scenario. Participants attributed most of their learning to exposure to teaching moments in clinical practice.
Conclusions: Engaging parents in a shared decision-making process remains a challenge for which there is little structured training. Consultants demonstrated an awareness of the value-laden nature

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

of this decision. This study indicates a role for optimising learning opportunities for trainees in paediatric cardiology both in teachable moments in practice, and in more formalised training focused on adult learning needs.

P-72
Prevalence and associated comorbidities of fetal ventricular septal defect (VSD)

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Introduction: Despite isolated VSD (iVSD) is the most common congenital heart disease in a neonate, still little is known on the topic in regards of fetal echocardiography. The aim is to evaluate iVSD in terms of frequency, as well as the associated morbidity of the fetus.
Methods: In retrospective analysis we identified 2005 confirmed cases of congenital heart disease diagnosed at tertiary referral center. The data was analyzed in regards of maternal characteristics, fetal extracardiac and chromosomal abnormalities. The location of VSD was defined using simplified echocardiographic classification as either perimembranous, muscular, inlet (not associated with AVSD), subarterial, and multiple or unclassified.
Results: There were 296 cases of iVSD, 14.7% of all CHD in the studied period. Fifty-two (17,6%) were classified as perimembranous, 51 (17.3%) as inlet, 42 (14.2%) as muscular, 13 (4.4%), 8 (2.7%) multiple VSD, with the rest unclassified (mostly in early series). Five most common reasons for referral were abnormal heart at ultrasound (61%), extracardiac abnormalities at ultrasound (12%), mothers age >35 years (11%), increased nuchal translucency (8%), chromosomal aberrations of the fetus (6%). The mean mothers age was 31 (18-44) years with correlation to the type of the defect (p>0,05).
Various extracardiac abnormalities were found in 108 (36,6%) of the fetuses, in some multiple, the results are presented in Table 1. One hundred forty-nine fetuses had genetical testing for various indications, the results are shown in Table 2. There was a strong association of inlet VSD and Edwards syndrome (p<0.001) as well as some extracardiac abnormalities (p=0.050 – p<0.001) as shown in the last column of the Tables 1. and 2.
Conclusions: In fetuses with isolated VSD there is a high prevalence of associated extracardiac and chromosomal abnormalities with the highest risk of comorbidity related to inlet VSD. This is crucial to consider when counseling parents and planning genetical testing.

Table 1. Extracardiac abnormalities

Extracardiac abnormalities	N, percent	Associated VSD type
Aortic arch and its branches	10, 3.4%	
Persistent left persistant SVC	9, 3.0%	
Central nervous system	50, 17.0%	Inlet VSD, p<0.001
Gastrointestinal	36, 12.2%	Inlet VSD, p=0.050
Craniofacial	26, 8.8%	Inlet VSD, p=0.014
Skeletal	25, 8.5%	Inlet VSD, p<0.001
Genito-urinary	24, 8.1%	Inlet VSD, p=0.044
Thorax	3, 1.0%	

Table 2. Chromosomal aberrations

Chromosomal aberration	N, % of tested, % of total	Associated VSD type
Edwards	48, 32%,16%	Inlet, p<0.001
Downs	12, 8%, 4%	Perimembranous, p<0.001
Patau	9, 6%, 3%	
Turner	2, 1%, 1%	
Other	11, 7%, 4%	
Normal	68, 46%, 23%	

P-73
Unusual Early Prenatal Presentation of Aicardi-Goutieres Syndrome

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Aicardi–Goutieres syndrome (AGS) is a rare genetic disease arising during the first year of life mainly affecting the central nervous system. Fetal AGS mimics in-utero infection and presents with microcephaly and cerebral calcifications.
We report on prenatal myocardial calcifications as the first manifestation of AGS. The parents are consanguineous with two healthy children and one who suffered from severe intellectual disability, epilepsy and diffuse brain calcifications on CT scan. He died at 4 years of age without a definitive diagnosis.
At 21 weeks of the current pregnancy, myocardial calcification with premature atrial beats and pericardial effusion were observed. No calcifications were detected in the brain, liver, spleen, or placenta. Maternal serology for intrauterine infection, ANA, anti-SSA and anti-SSB were negative. Ultrasound at 27 weeks revealed calcifications of the myocardium, caudothalamic groove and liver. At 32 weeks, reduced cardiac contractility, multiple cardiac, thalamic, brainstem and striatal calcifications and a parenchymal temporal lobe cyst were detected.
The couple chose to terminate the pregnancy. Genetic analysis of the amniotic fluid revealed biallelic mutations in TREX1 gene compatible with AGS. Notably, this same mutation was detected in the preserved DNA of the deceased sibling.
AGS is associated with a high risk of recurrence. Since AGS calcifications may mimic in-utero infection, AGS should be considered when TORCH is negative. Cardiac involvement with abnormal contractility, calcifications, effusion and arrhythmia are unusual in AGS but may be the earliest manifestation of AGS due to TREX1 mutation.

P-74
Total anomalous pulmonary venous connection and the nutmeg lung pattern in a fetus: prognostic indicator for counseling and outcome?

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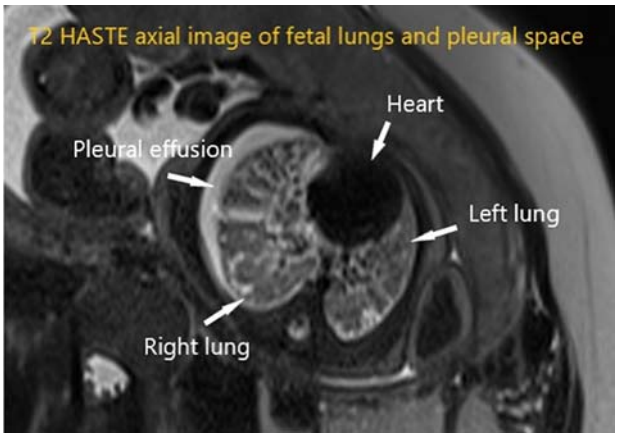
Background: The term nutmeg lung has been used to describe the heterogeneous appearance of the lung parenchyma of primary or secondary congenital lymphangiectasia. We describe the nutmeg lung appearance seen in a fetal magnetic resonance imaging (MRI) scan in a fetus with pulmonary venous hypoplasia, ventricular septal defect and hypoplastic aortic arch with an adverse outcome, with special focus on the prognostic role of MRI in counseling and preparation for delivery.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



Methods: The prenatal ultrasound and MRI findings of a fetus with complex congenital heart disease and congenital lymphangiectasia were reviewed and correlated with postnatal imaging findings and clinical outcome.
Results: A male fetus was referred for a cardiac evaluation at 35 and 4/7 weeks gestational age (GA) due to bilateral effusions and hypoechoic lung parenchyma. The echocardiogram revealed total anomalous pulmonary venous connection (TAPVC) with possible pulmonary venous hypoplasia, ventricular size discrepancy with right ventricular dominance and a mildly hypoplastic aortic arch. Significant bilateral pleural effusions and a small pericardial effusion, as well as abnormal echogenicity of the lung parenchyma were also identified. Subsequent fetal MRI consisting mainly of T2 weighted sequences, revealed abnormal lung parenchyma with pulmonary lymphangiectasis with grossly dilated lymphatic channels in the subpleural, interlobar, perivascular, and peribronchial areas as well as bilateral pleural effusions. Induction of labor was performed at 38 weeks (GA) with anticipation for urgent need for catheterization and possible ECMO (extracorporeal membrane oxygenation). Immediately after birth hemodynamic and respiratory instability developed and the pulmonary venous anatomy could not be imaged echocardiographically. Due to severe cyanosis and hemodynamic instability the newborn was placed on ECMO; subsequent catheterization revealed (TAPVC) with diffuse pulmonary venous hypoplasia, absence of a decompressing vein and coarctation of the aorta. The cardiac defects were deemed inoperable by cardiology and cardiac surgery and the parents chose palliative care; the newborn expired at 27 hours of life.
Conclusions: Presence of a nutmeg lung in utero may result from secondary pulmonary lymphangiectasia due to significant congenital heart defects such as TAPVC and hypoplasia. Its identification is essential for prenatal counseling and delivery planning, frequently involving EXIT to ECMO and catheterization laboratory.

P-75
Fetal left ventricular strain – Impact of angle of insonation and frame rate

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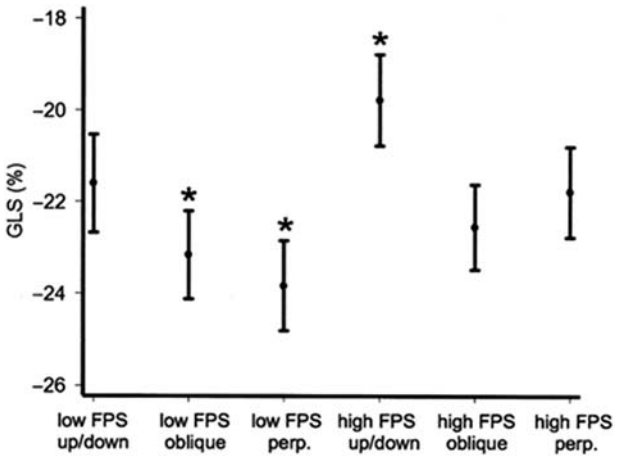

Figure.
*perp., perpendicular, *p <0.05.*

Introduction: Speckle tracking echocardiography (STE) is used increasingly as a method to assess fetal myocardial deformation and velocity in health and disease. However, comparison of results among different studies is challenging as temporal resolution and position of fetal heart relative to the angle of insonation differs. The primary aim of this study was to explore whether temporal resolution (frames per second, FPS) and angle of insonation (apex position) can influence left ventricular endocardial global longitudinal strain (GLS), and the secondary aim was to report the reproducibility of the analysis of fetal STE.
Methods: Ultrasound clips of the fetal four-chamber view were obtained during routine clinical scanning in 75 healthy fetuses from 19-38 weeks gestation, using Canon Aplio i800 machines. The analysis was performed by two trained operators (JS, TD) using Vitrea software (Canon) to calculate left ventricular GLS. For every fetus, three different orientations of the fetal heart (apex up/down, apex perpendicular, apex oblique) were obtained, at low and high acoustic FPS. Analysis was performed using linear mixed model analysis. The intra and inter-analyser reproducibility was evaluated in 40 clips which were analysed by both operators blinded to the other's findings
Results: Analysis was performed on 312 clips (144 in 2nd trimester; 168 in 3rd trimester). FPS and angle of insonation were important determinants of GLS. Higher FPS (mean 118.4 ± 34.4), compared to lower FPS (62.6 ± 13.5), for the same scanning site resulted in lower GLS (-19.6% ± 2.8 vs. -21.3% ± 4.0, $p<0.001$). Apex perpendicular views were associated with higher GLS in comparison with apex up/down (-23.6% ± 4.1 vs. -21.3 ± 4.0, $p<0.001$). The composite influence of FPS and position of fetal heart on GLS is illustrated below (figure). There was good intra and inter-analyser reliability of GLS (intraclass correlation coefficient 0.88 and 0.84 respectively).
Conclusions: Our results indicate that the angle of insonation and acquisition frame rate are important determinants of GLS. These factors should be taken into account when comparing studies using different protocols of acquisition. Speckle tracking cannot be regarded as an "angle independent" modality during fetal life.

P-76
Impact Of Gender On Outcomes Of Congenital Heart Disease In Childhood: a 21 year experience of a prenatally diagnosed cohort

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Background: Variations in the presentation and outcome of heart disease according to sex are well-described. Our objective was to report the impact of gender on outcome of congenital heart disease in a cohort of prenatally diagnosed lesions.
Methods: Cases with a prenatal diagnosis of either: classical hypoplastic left heart syndrome (HLHS), tetralogy of Fallot (TOF), isolated transposition of the great arteries (TGA), coarctation of the aorta, balanced atrioventricular septal defect (AVSD) or heterotaxy born between 1995 to 2016 were identified from the fetal cardiac database. Postnatal medical records were reviewed; information sought on associated anomalies, genetic associations, cardiac surgeries, complications and survival assessed according to sex.
Results: There were 990 babies fulfilling the inclusion criteria of which 397 (40.1%) were female. Coarctation of the aorta, TGA and HLHS were more common in males. The overall actuarial survival was 74% (732/990) with the majority of deaths occurring in the early surgical period. In survivors the median time of follow-up was 102 months (range: 0.2 - 281 months). Four patients were underwent cardiac transplantation. There was no significant difference in mortality (p=0.9), cause of death (p=0.2), and complications (p=0.8) according to sex.
Re-intervention of the primary cardiac procedure were as follows: re-coarctation: 10/182 (5.5%; 5 female), AVSD: 14/138 (10.1%; 7 female), TGA: 10/147 (6.8%; 4 female) and TOF: 30/210 (14.3%; 16 female). There were 12/210 (5.7%) pulmonary valve replacements (4 female) in patients with repaired TOF. Secondary operations on patients with HLHS were performed in 55/232 (23.7%): tricuspid valve repair: 21 (9%; 8 female), branch pulmonary artery 17 (7.3%; 7 female) and re-coarctation 17 (7.3%; 6 female). There was no significant difference in the frequency of re-intervention according to sex (p=0.4).
Conclusions: Medium term outcome and survival into early adulthood does not vary between males and females in the current era. Although congenital heart disease has been associated with significant morbidity and mortality in adulthood it is hoped that improvements in the paediatric journey will translate into better survival and quality of survival in adulthood and services will be able to support the distinct issues faced by both female and male adults.

P-77
Impact of antenatal diagnosis on outcomes after arterial switch operation for transposition of the great arteries in children

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The objective of this study was to assess the long term outcome of children after arterial switch operation (ASO) for transposition of the great arteries (TGA) and compare patients with and without antenatal diagnosis.
Material and methods: retrospective analysis of all APS performed from 2009 to 2016 in patients with unrepaired cyanotic CHD. Demographics, clinical and biological data, surgical techniques, echocardiographic measurements, and outcomes were collected. Patients with antenatal diagnosis (AN group) and those diagnosed after birth (PN group) were compared.
Results: 364 patients were included (263 males): 248 without VSD (68%) and 116 with one small VSD (32%). Antenatal diagnosis was made in 60,4% of the cases (37.6% < 2008, 76.5% > 2008). AN group included 220 patients and PN group 144 patients. Birth weight was 3207±511g, gestational age was 39.1± 1.6weeks (38.±1.6w in AN vs 39.5±1.5 in PN groups, p= 0.0016), birth weight was similar. Age at diagnosis was 2.3±6.2days in PN group. Preoperative ventilatory and prostaglandins support were less frequent in AN than PN (respectively 34% vs 82%, et 44% vs 70%, p< 0.0001). Time to Rashkind procedure after birth was 0.03±0.1 in AN vs 1.1±2.8days in PN, p <0.0001. Age at surgery did not differ between AN and PN groups: 7.2±6 vs 7.7±6.5days, p= 0.504. Long term follow-up was 9.2±4.8 years. Survival rates were 95.3% at 1 month, 94.8% at 3, 6 and 12 months and 94.5% at 2.5 years and up to 18 years after surgery. Twenty patients died (5.5%): 17 early postoperative (4.7%). Time to death was 64.2 ±207.9days (0 to 919). Mortality was similar between both groups: 5% in AN vs 6.3% in PN, p= 0.609.
Conclusion: The results of his study showed that gestational age a birth was lower and time to Rashking procedure was shorter in neonates with TGA diagnosed antenatally than postnatally but age at surgery and outcomes were similar.

P-78
Prospective study for antenatal diagnosis of coarctation of the aorta

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The objective of this study was to define echocardiographic (ECHO) parameters during fetal life, to predict postnatal aortic coarctation (COA).
Material and Methods: This prospective single-center study from 2010 to 2018 included all fetus diagnosed with isolated ventricular and/ or great vessels asymetry (right heart dominance). Complex CHD were excluded. The cohort was divided in patients with coarctation after birth (COA) and those free from coarctation (noCOA). Left heart, aortic and ductus measurements were collected serially at second trimester (T2), third trimester (T3) ad Z-scores were assessed. COA and noCOA were compared, sensitivity (Se) and specificity (Spec) and ROC curves (cut-off values) were assessed for each parameter.
Results: 67 cases were included: 34 (50.7%) ranged in group COA and 33 (49.3%) in noCOA. Coarctation occurred more frequently if suspected at T2 (70.6% in COA vs 30.3% in noCOA, p= 0.001). Mitral annulus (T3) was 8.6mm in COA vs 10.3mm in noCOA (p= 0.002) with cut-off < 7.3mm (Se= 50%, Spec= 93.6%, OR= 14.5). Aortic annulus diameter (T3) was 4.8mm in COA vs 6mm in noCOA (p= 0.005), with cut-off < 5.4mm (Se= 65%, Spec= 77%, OR= 6.3). Aortic isthmus diameter (T3) was 1.4mm in COA vs 2.8mm in noCOA (p= 0.003), with cut-off < 3mm (Se= 91%, Spec= 65%, OR= 18.9). Ductus arteriosus/ aortic isthmus ratio (T3) was 2.5 in COA vs 1.8 in noCOA (p= 0.01) with cut-off > 1.57mm (Se= 90.5%, Spec= 62%, OR= 15.4).
Conclusion: Mitral annulus < 7.3mm, aortic annulus < 5.4mm, aortic isthmus < 3mm and ductus/isthmus ratio > 1.57 assessed during 3rd trimester of fetal life may help to predict postnatal coractation of the aorta. These results have to be confirmed by larger prospective studies.

P-79
Prenatally diagnosis and outcome of fetuses with rhabdomyoma – single centre experience

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Background: Cardiac rhabdomyomas (CRs) are the most common primary tumors of the heart in neonates and infants/children. Usually are multiple and, based on the location, can cause haemodynamic disturbance, dysrhythmias or heart failure during the fetal and early postnatal period. CRs have a natural history of spontaneous regression and are closely associated with tuberous sclerosis complex (TSC). It has an association with tuberous sclerosis (TS) and in those cases the tumor may regress and disappear completely, or remain consistent in size.
Objective: We aimed to evaluate the prenatal diagnosis, clinical presentation and outcome of CRs and their association with TSC in a single centre. The median follow-up period was three years (range: 6 months - 5 years).
Methods: We reviewed medical records of all foetuses diagnosed prenatally with cardiac rhabdomyomas, covering the period from January 2010 to December 2016, which had undergone detailed ultrasound evaluation at a tertiary level centre with limited technical resources.
Results: Twelve fetuses were included in the study; all with multiple tumors and, a total of 53 tumors were identified in all patients - the maximum was one fetus with 12 tumors. All patients were diagnosed prenatally by fetal echocardiography. In two foetuses haemodynamic disturbances during the fetal period were noted and pregnancies have been terminated. After long consultation, termination of pregnancy was chosen by the parents in totally 8 cases. In four continuing pregnancies, during the first year of life tumors regressed. TSC was diagnosed in all patients during the follow-up.
Conclusions: Cardiac rhabdomyomas are benign from the cardiovascular standpoint in most affected fetuses. An early prenatal diagnosis may help for an adequate planning of perinatal monitoring and treatment with involvement of a multidisciplinary team. Large tumor size, number of tumors and localization may cause hydrops and they are significantly associated with poor neonatal outcome.

Keywords: heart tumor, cardiac rhabdomyoma, fetal echocardiography, tuberous sclerosis complex.

P-80

Utility of fetal diagnosis of congenital heart disease in the Japanese countryside

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Objectives: To evaluate the effectiveness of fetal diagnosis of congenital heart disease (CHD) in Japanese countryside which has a limited number of institutions in which pediatric cardiac surgery can be performed.
Methods: We retrospectively examined 202 neonates referred to our institution for cardiac surgery within 14 days after birth. Subjects were divided into groups A (n=44) and B (n=158), before and after the establishment of the fetal echocardiography outpatient clinic (FEOC) in our institution, respectively. We evaluated the fetal diagnosis rate, emergency hospitalization rate, inhospital mortality, and cardiac operative risk evaluation score of each group.
Results: The fetal diagnosis rate of CHD was 20.5% in group A and 46.8% in group B. Group B had a significantly lower emergency hospitalization rate (22.7% vs 8.9%, p=0.018).There is no significant difference in in-hospital mortality between the groups (13.6% vs 13.9%), despite group B had a significantly higher cardiac operative risk evaluation score (1.03 ± 0.10 vs 1.43 ± 0.05, p=0.0004).
Conclusions: Fetal diagnosis rate of CHD had been increased after the establishment of FEOC. Fetal diagnosis of CHD is effective for decreasing the emergency hospitalization rate and inhospital mortality of neonates who need high risk cardiac surgery.

P-81

Repeated rapid regression of two cardiac rhabdomyoma in a newborn with Everolimus

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Introduction: Cardiac rhabdomyoma (CRM) occur in 50% of patients with tuberous sclerosis complex (TSC). CRM are frequently asymptomatic and tend to regress spontaneously. However, some CRM cause arrhythmias, obstructions or valve movements disorders. Current treatment of choice in these patients is surgical resection. Recently encouraging experiences with mTOR-inhibitors such as Everolimus have been reported.
Case Description: A giant left ventricle tumor was detected antenatally with ultrasound in a fetus. The tumor was considered to be a rhabdomyoma, the most common heart tumor in infants. The child was born on term and showed good primary postnatal adaptation. Two Ash-Leaf spots on the right forearm indicated the presence of TSC, which made the diagnosis of CRM even more likely. Prostaglandin E1 was administered from the first hour of life. Echocardiography after birth showed a large subaortal CRM (10x6x8mm) with subtotal obstruction of the LVOT (supravalvular gradient 70mmHg) and a second septal right ventricular CRM (22x13x10mm). Surgical therapy was rejected due to the high risk of aortic valve destruction. Thus, oral therapy with Everolimus with 0.03mg/kg/d and a target serum trough level of 5-8ug/l was initiated on the third day of life.
Echocardiography showed a volume regression of more than 80% in both CRMs within six weeks. There was also no longer any supravalvular gradient. Everolimus was therefore ceased 3 months after birth to evaluate whether further regression will occur spontaneously. Four months later follow-up echocardiography revealed regrowth of both CRMs and increasing LVOT obstruction without clinical symptoms. Immediate restart of Everolimus resulted once again in rapid regression of both CRMs and LVOT obstruction was no longer measurable after four weeks. Regular follow-up echocardiographies confirmed continuing shrinkage of both CRMs even though they are still present at the age of 19 months. Due to TSC-associated intractable seizures Everolimus is still administered.
Conclusion: Beside established indications Everolimus may be a promising alternative for high-risk surgical cases with hemodynamically significant CRM. As shown in our patient, CRM can respond repeatedly which offers the opportunity of early treatment cessation awaiting spontaneous regression as well as avoiding therapy-associated side effects.

P-82

Assessment of cardiovascular profile score in fetuses with agenesis of ductus venosus, without structural heart defect.

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Introduction: The agenesis of ductus venosus (ADV) is a rare abnormality, which is divided into two types, intrahepatic and extrahepatic. That may cause functional abnormalities in the

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

circulatory system. The ADV is associated with the higher risk of the heart defects, chromosomal anomalies and congestive heart failure.
Methods: Forty four fetuses with the diagnosis of ADV were analyzed retrospectively in referral center between April 2016 and December 2018.
Nineteen of them with anatomically normal heart (19/44 - 43%) were included to the final analysis. ADV has been divided into intrahepatic and extrahepatic type and analyzed separately.
Cardiovascular profile score (CVPS) was assessed in all group and diameter of the shunt (portosystemic) was assessed in group with extrahepatic type of ADV.
Results: Extrahepatic type of ADV was recognized in 4/19 (21%) cases and all of them presented functional changes in cardiovascular system. In 3/4 cases (75%) occurred cardiomegaly, 2/4 (50%) moderate tricuspid regurgitation, in 1/4 (25%) fetal hydrops and in 1/4 (25%) extension of inferior vena cava. Average CVPS was 8 points. Shunt was wide in all cases of extrahepatic ADV.
Intrahepatic type of ADV was found in 15/19 cases (79%) and 9 (47%) of them presented mild tricuspid regurgitation. Average CVPS was 10 points.
Conclusions: Agenesis of ductus venosus with no structural heart defect occured more frequently in intrahepatic type than in extrahepatic.
All fetuses with extrahepatic ADV type presented functional changes in circulatory system in opposite to those with intrahepatic type where, CVPS was 10 points.
The most common functional abnormality in group of extrahepatic type ADV was cardiomegaly.

P-83
Factors associated with exercise capacity in patients with a systemic right ventricle

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Background: Systemic right ventricle (SRV) is a rare and complex congenital heart disease (CHD). Patients with SVR present a significant decrease of their exercise capacity but the prediction of this decline in clinical practice is still challenging.
Aims: We aimed at identifying clinical and paraclinical factors associated with maximum oxygen uptake (VO2max) in adults with SRV.
Methods: We performed a multicentre cross-sectional study from January to December 2017 in three French tertiary CHD centres. All adults with D-transposition of the great arteries (d-TGA) and congenitally corrected TGA (cc-TGA) were included. Demographic, clinical, laboratory and imaging data were collected. Univariate and mulitvariate analyses were performed to identify predictors of impaired VO2max, as measured by cardiopulmonary exercise test (CPET).
Results: A total of 111 patients were included in the study (85% d-TGA, median age 37.2 ± 8.2 years). Nearly 2/3 (n=70) of the patients presented with various conduction disorders and 1/3 (n=34) with cardiac rhythm disorders. Median B-type natriuretic peptide (BNP) was 58.5 [10-995] pg/ml. All echocardiographic parameters showed at least a mild SRV dysfunction and 17% of the patients presented with severe tricuspid valve regurgitation. VO2max was impaired in all patients (mean 23.3 ± 6.9 ml/kg/min, representing 42.0 ± 7.4% of predicted values). In univariate analysis, NYHA functional class, BNP levels, right ventricular dysfunction, severity of tricuspid valve regurgitation, and the presence of a pacemaker and/or an implantable defibrillator, were correlated with VO2max. In multivariate analysis, only the patient's self-assessment of functional status, as measured by NYHA functional class was correlated with VO2max (P=0.005).
Conclusion: NYHA functional class is the strongest predictor of impaired exercise capacity in adult patients with SRV.

Keywords: systemic right ventricle, cardiopulmonary exercise test, NYHA functional class, congenital heart disease.

P-84
The role of NT-proBNP in clinical follow-up of patients with repaired Tetralogy of Fallot

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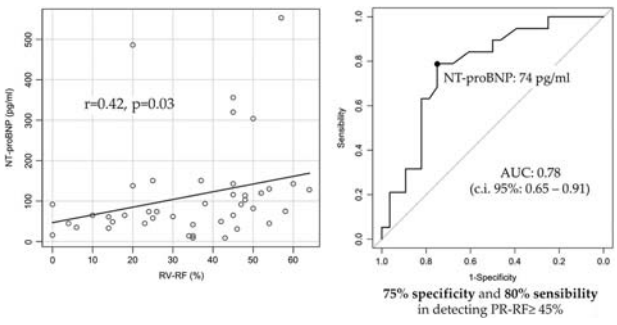
Introduction: Patients with repaired TOF often develop right ventricle (RV) dilation and dysfunction due to pulmonary regurgitation (PR) or RV outflow tract (RVOT) obstruction and later need pulmonary valve replacement (PVR).
Methods: In order to demonstrate a correlation of NT-proBNP concentrations with clinical status and exercise capacity, we consecutively enrolled repaired TOF patients at the time of a scheduled periodic follow-up visit. All patients underwent complete physical examination, blood testing including NT-proBNP, 12-lead ECG, ambulatory ECG, cardiopulmonary exercise test (CPET) and echocardiography. R statistical software was used for analysis. Since numeric variables were not normally distributed, non-parametric tests were performed.
Results: From September 2017 to October 2018, a total of 51 patients were enrolled. Baseline characteristics of study population are shown in Table 1. NYHA functional class was associated with residual intra-cardiac defects ($\chi 2$, p=0.002), mitral regurgitation ($\chi 2$, p=0.008), and the occurrence of bradyarrhythmias ($\chi 2$, p=0.0001), polymorphic PVCs ($\chi 2$, p=0.04) and non-sustained ventricular tachycardia ($\chi 2$, p<0.0001) at 24h ambulatory ECG. Median NT-proBNP value was 74.0 pg/ml (IQR 44.25 – 130.75; range 8.09 – 2224.0). It showed a significant association with NYHA functional class (Kruskal-Wallis, p=0.002). NT-proBNP levels were associated with signs of right chambers enlargement, as they were positively correlated with distal RVOT diameter (r=0.27, p=0.05) and to RA area z-score (r=0.65, p=0.0001). Patients with significant PR had significantly higher NT-proBNP values (Mann-Whitney, p=0.004). A positive significant correlation was found between NT-proBNP and PR regurgitant fraction (PR-RF) (r=0.42, p=0.03). NT-proBNP demonstrated good accuracy in predicting the presence of a severe PR (PR-RF≥ 45%): area under the ROC curve was 0.78 (c.i. 95%: 0.65 – 0.91). The best cut-off value was 74.5 pg/ml (75% specificity and 80% sensibility). None of the main CPET parameters had a significant correlation to NT-proBNP. However, the biomarker



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Age –y	15.1 (10.9 – 21.65)
Male sex – no. (%)	32 (62)
mBT shunt before repair – no. (%)	18 (35.3)
Age at repair - mo	13 (6 – 24)
Type of repair – no. (%)	
Trans-annular patch	30 (58.8)
Infundibular patch	15 (29.4)
RV-to-PA conduit	5 (9.8)
Conservative (trans-pulmonary)	1 (2)
NYHA class – no. (%)	
I	34 (66.7)
II	16 (31.3)
III	1 (2)

was positively correlated to the level of desaturation (basal SpO_2 – peak SpO_2) during CPET (r=-0.35, p=0.02)
Conclusions: NYHA class was associated to the occurrence of bradyarrhythmias and ventricular tachyarrhythmias and with NT-proBNP levels, which were strongly correlated to right chambers enlargement and consistently to PR severity. NT-proBNP determination could contribute to establish the timing of PVR in repaired TOF patients with severe PR.

P-85
Sexual dysfunction in male adult patients with CHD

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Background: Little is known about the impact of the CHD on sex life. At the same time, the few information available rarely reach those affected. Whether and to what extent sex life is discussed with the treating physician is unknown.
Purpose: To study the knowledge level, problems, wishes and fears of male ACHD regarding their sex life.
Methods: Patients recruited via the German National Register for Congenital Heart Defects were invited to an online survey about sex life. 2,221 male patients were invited to the survey and 371 participated (16.7%). Patients were divided into four groups based on the CHD severity classification of Warnes et al.: (A) simple CHD (40 patients), (B) moderate CHD (157 patients), (C) complex CHD (152 patients) and (D) other CHD (22 patients).
Results: 132 (35.6%) patients reported that they had very rarely or no sex at all in the last six months (A: 27.5%, B: 35%, C: 37.5%, D: 40.9%). Based on the last six months, 71.2% of the surveyed patients estimated the probability of having a maintaining erection as high or very high (A: 85%, B: 71.4%, C: 69.7%, D: 54.5%). In the last six months, 60.6% of the interviewed patients, according to their own information, were almost always or always able to get a full erection sufficient for the sexual intercourse (A: 77.5%, B: 62.4%, C: 55.3%, D: 54.5%). 14.6% already had erectile dysfunction, but did not discuss this with their physician (A: 10%, B: 14.6%, C: 15.1%, D: 18.2%). Overall only 3.5% of the questioned patients stated that they were offered a consultation regarding erectile dysfunction by their treating physician. However, 29.6% would like to receive such counseling during routine medical examinations.
Conclusions: This study reveals important issues regarding sex life and erectile dysfunction in male ACHD. Additionally there were differences found regarding the CHD severity. A discussion about sex life should take place with every ACHD patient during regular clinical contacts to find solutions for possible problems regarding sex life.

P-86
Long-term follow-up in patients after Tetralogy of Fallot repair: 20 years single-centre experience

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Introduction: Tetralogy of Fallot (TOF) is the most common cyanotic heart defect with long-term survival after complete correction. The aim of our single-centre whole-country study was to evaluate results and late complications after complete correction.
Methods: Performed was retrospective analysis of TOF patients, who underwent correction at our institution in the period 1992-2002 and were at least 15 years after complete surgical repair. Included were 181 patients, median follow-up 21.8 years (16-25 years). Serial complete cardiology examinations were performed.
Results: Overall freedom from re-do was 64.6%.
Early reinterventions were needed in 23 patients (12,7%): residual shunt closure in 4, right ventricular (RV) outflow tract obstruction relief in 8, pulmonary artery branch balloon dilatation and/or stent implantation in 11 patients; median time 3.6 years (8 months-4.8 years) post complete correction.
Late reoperation (pulmonary conduit implantation) was performed in 22 patients (12.2%), with median of 16.7 years (8.6-20.5 years) after complete correction.
Clinically relevant arrhythmias were found in 29 patients (16.1%); 13 patients (7.2%) needed long-term anti-arrhythmic medication, 4 electrical cardioversion, and 1 an ICD implantation.
Transannular patch (TAP) was a significant risk factor for: severe pulmonary regurgitation with RV dilatation (p=0.0002), for pulmonary conduit implantation (p<0.0001), as well as for the presence of hemodynamically considerable ventricular arrhythmias (p=0.008). TAP with secondary right atrial dilatation correlated also with the presence of severe atrial arrhythmias (p=0.036).
In patients with isolated RV outflow tract aneurysm significantly lower global RV function (as measured by MRI) was found (p=0.027). In 71.4% of these patients, the decreased RV function (EF<40%) established by MRI, was not confirmed by echocardiography.
Conclusions: Clinical findings in TOF patients after definitive surgical correction are very good. There is a low occurrence of late complications throughout childhood, although this increases with age, especially when reaching young adulthood. TAP is the most significant risk factor for RV dilatation and with the secondary need of conduit implantation, as well as for the presence of severe arrhythmias. Isolated RV outflow tract aneurysm is an important finding that has to be taken into account as it can lead to significant underestimation of RV function measured by MRI.

P-87
Percutaneous edge-to-edge Repair of the Systemic AV Valve with the MitraClip System to Preserve Valve Function in Congenitally Corrected Transposition

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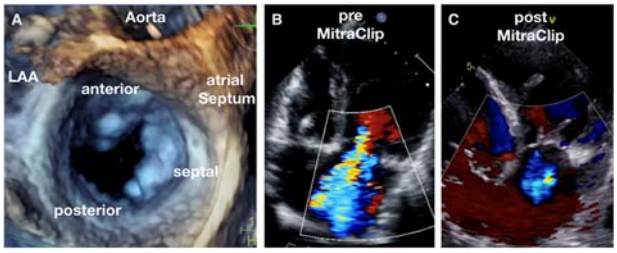
https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



Objectives: Clinically significant regurgitation of the systemic AV valve (SAVVR) is a common occurrence in congenital corrected transposition (ccTGA). Percutaneous edge-to-edge repair with the MitraClip system might be a viable option to treat this problem.
Methods: We describe two female patients, aged 62 and 72 years respectively with simple ccTGA and no history of previous cardiac surgery. Both presented with severe (grade 4) SAVV on transthoracic (TTE) and transesophageal (TEE) echocardiography associated with severe dilatation of the systemic ventricle. Both were in NYHA functional class III. 3D-TEE revealed an additional prolapse of the anterior leaflet in one patient. Both underwent MitraClip (Abbott, USA) repair, after informed consent was obtained, as an alternative to surgery.
Results: Procedures were performed under general anaesthesia, with TEE guidance. Via a trans-septal approach, one MitraClip was successfully deployed by grasping the septal and anterior leaflets in each case. Post-procedural SAVVR was reduced to grade 1 in both patients, with no stenotic component. Both patients demonstrated clinical improvement, to NYHA class 1. On serial echocardiographic follow-up up to 3 months SAVVR remained at grade 1 in patient 1, and at grade 2 in patient 2.
Conclusions: Percutaneous edge-to-edge repair with the MitraClip system is a valuable, minimally invasive option to treat severe SAVVR in selected patients with ccTGA.

P-88
Prognostic index to evaluate the risk of the right ventricle dysfunction faced with systemic afterload

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The introduction of physiological atrial correction surgery allowed prolonging the life expectancy of those born with transposition of large arteries. In Cuba, no index is applied to assess the risk of right ventricle dysfunction faced with systemic afterload.
Objective: The objective of this research was to develop and validate a prognostic index for this purpose.
Method: An observational, prospective, cross-sectional study was conducted in 90 patients of the Pediatric Cardiocenter "William Soler" from 2012 to 2015. A logistic regression model was performed to identify the variables that contributed to significant independent risk of dysfunction. To determine the clinical relevance of the echocardiographic variables, an effectiveness study was carried out with analysis of the incidence and prevalence of the event (clinical dysfunction of the right ventricle facing systemic afterload), at each echocardiographic measurement.
Results: Initial systolic dysfunction of right ventricle faced with systemic afterload was observed, dependent on the increase in afterload and not due to myocardial disorder. There are no alterations in the initial diastolic function. The variables that make up the prognostic index are: end-diastolic, end-systolic and parietal thickness of the right ventricle, systolic excursion of the tricuspid annular plane, ejection fraction of the right ventricle, S wave, Tei index, pressure derivative as a function of time and characterization of tricuspid regurgitation.
Conclusions: The index shows good discriminatory capacity and adequate calibration in prediction of ventricular dysfunction. Telediastolic diameter and parietal thickness of the right ventricle, derived from pressure as a function of time and characterization of tricuspid regurgitation, demonstrate clinical relevance. The prognostic index shows validity and allows its introduction in clinical practice.

P-89
Metabolomic profiling as a promising tool to identify biomarkers in Fontan patients

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Objectives: Metabolomics is the study of small organic molecules processed along biochemical pathways. Using this technique to analyse minute biospecimens, novel candidate biomarkers for heart failure have been identified. Up to now little is known about metabolism in Fontan patients. We aimed to analyse the metabolic pattern in adult Fontan patients with a left dominant ventricle.
Methods: We determined the metabolic pattern of 20 adult Fontan patients with a dominant left ventricle, and 20 age and gender matched healthy biventricular controls, using a 0,5 ml serum sample each and the BIOCRATES AbsoluteIDQ p180 kit. Between groups we compared metabolite concentrations of the protein and lipid metabolism.
Results: Compared to biventricular controls, in Fontan patients serum phosphatidylcholines and sphingomyelins were significantly decreased ($q < 0.05$), while the modified proteins methionine-sulfoxide (Met-SO, $q < 0.001$) and asymmetric dimethylarginine (ADMA, $q = 0.0018$) were significantly increased.
Conclusions: There is a distinct metabolic pattern indicating structural membrane alterations (decreased phosphatidylcholines and sphingomyelins), oxidative stress (increased Met-SO), and alteration in NO-dependent signalling processes (increased ADMA) in Fontan patients with a systemic left ventricle. Requiring minute amounts of biospecimens to analyse multiple metabolic pathways simultaneously, metabolomic profiling is useful for the assessment of metabolic derangement, possibly delivering a promising tool to identify biomarkers for the Fontan circulation.

P-90
Successful surgical management for cyanotic congenital heart disease complicated with pulmonary aspergillosis. - A case report

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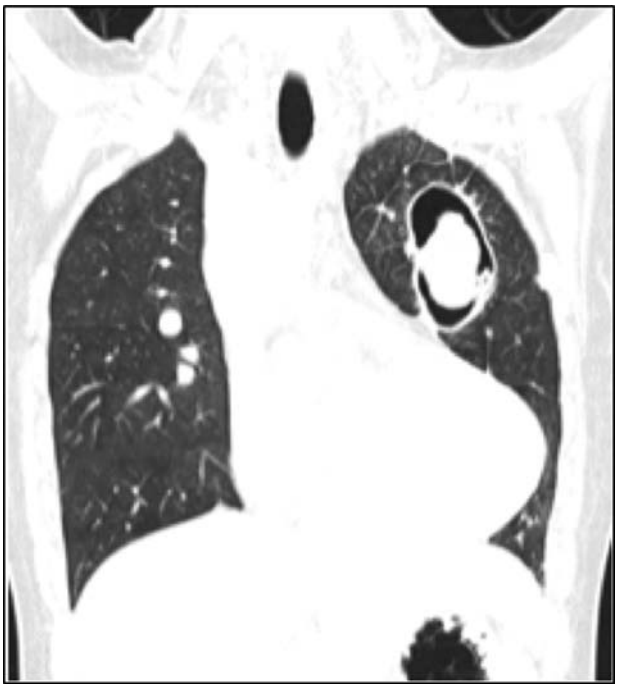
https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



Introduction: Pulmonary aspergillosis complicated with cyanotic congenital heart disease is rare, but known to have a quite poor prognosis. We experienced a successful surgical case with truncus arteriosus and major aortopulmonary collateral artery (MAPCA) who was performed primary Rastelli procedure after thoracoscopic lobectomy for progressed pulmonary aspergillosis.
Case Report: A 17-year-old female with naïve truncus arteriosus and MAPCA had repetitive hemoptysis because of progressed left pulmonary aspergillosis that was refractory to antibiotics therapy. Cardiac catheterization found developed collateral vessels around the cavity lesion of aspergillosis associated with poor pulmonary perfusion area, and also an indication of primary Rastelli procedure with developed bilateral pulmonary vascular bed without pulmonary hypertension. After coil embolization on collateral vessels, left upper lobectomy was performed under thoracoscopic surgery, and primary Rastelli procedure with unifocalization of MAPCA was performed 2 weeks later. Postoperative course was good without cyanosis, pulmonary hypertension and the recurrence of pulmonary aspergillosis.
Conclusions: This is a first report of successful surgical management for pulmonary aspergillosis complicated with cyanotic congenital heart disease.

P-91
Quality of life of children after surgical treatment of aortic coarctation

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Introduction: children and teens after surgical treatment (coarctation of the aorta) make up a special group of patients (grown-up congenital heart disease) (GUCH), who need constant supervision and specialized help with the aim of the complete restoration and preservation of health over the years. The quality of life is an integral characteristic feature of the physiological, psychological and social functioning of a healthy and sick person, which is based on their subjective perception. Data about the quality of life of children and adolescents after correction of aortic coarctation remain rather contradictory up to the present time.
Methods: Pediatric Quality of Life Inventory ™ Cardiac Module 3.0 questionnaire (PedsQL Cardiac Module). 56 children after surgical treatment of aortic coarctation took part: boys – 42, girls – 14. Age between 10 and 15 years old.
Results: 37 patients (66.07%) considered their lives to be full, 26.79% (15 people) – inferior, 7.14% (4 children) found it difficult to answer. All the patients reported that their physical activity was limited by doctors and parents. 25 (44.64%) of respondents pointed out the presence of communication problems and psychological difficulties; 42 people (75.00%) noticed hyperguardianship on the part of their parents. 16.07% (9 patients) suffered from arterial hypertension, in connection with this fact the use of antihypertensive drugs, constant monitoring of blood pressure, certain instrumental examinations with a prescribed frequency were necessary. All the above-mentioned factors created psychological difficulties for patients; 15 (26.79%) respondents complained of the deterioration of well-being in the form of cardialgia, headaches, poor exercise tolerance; 10 (17.85%) patients experienced learning difficulties preconditioned by the congenital heart disease.
Conclusions: despite the satisfactory results of the surgical treatment, patients showed a decrease in the quality of life owing to various factors. The most significant were: deteriorating quality of life, limited physical activity, constant monitoring by parents, obtaining medical treatment and the necessity for regular consultations with doctors.

P-92
Bicuspid Aortic Valve and Dilated Cardiomyophaty: two separate entities or common genetic link?

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Introduction: Whilst the genetic association between bicuspid aortic valve (BAV) and hypertrophic cardiomyopathy (HCM) has been reported in the literature, very little is known about the coexistence of BAV and dilated cardiomyophaty (DCM). This study investigates the prevalence of coexistent DCM in a BAV population.
Methods: Retrospective data of 196 consecutive BAV patients who attended the Cardiac Outpatients Clinics between June 2012 and October 2018, were analyzed. Data included assessment of family history of inherited cardiac conditions and congenital heart diseases, ECG, transthoracic echo and cardiac magnetic resonance imaging (CMR).
Results: Out of 196 patients, 179 patients were included since free from other associated cardiac lesions which could have caused ventricular volume overload such as ventricular septal defect and patent ductus arteriosus. Mean age was 34.6±13.5, male 128 (71%). 111 patients had isolated BAV, whilst 68 patients had BAV with aortic coarctation (CoA). 43 patients required surgical or transcatheter procedures for aortic valve (AV) dysfunction (mean age 26.1±11.1), 37 patients had CoA repair or stent (mean age 11.6 ±10.4). In patients who were free from previous AV procedures (n=136), there was evidence of mild AV stenosis (peak velocity 2.1±0.9 m/s) and/or regurgitation (regurgitant fraction 9±12%). Of these, 25(19%) had evidence of left ventricular (28%), right ventricular (12%) or both left and right ventricular dilatation (60%) which could not be explained by the degree of AV dysfunction or other right sided lesions for the RV. In particular, 14 patients had ascertained DCM and were under specialist follow-up. On CMR, their left ventricular end-diastolic volume

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

was 124±24ml/m^2, right ventricular end-diastolic volume 110± 25ml/m_2, left ventricle ejection fraction 55±8% and right ventricle ejection fraction 57±6%. In 11 patients DCM was suspected but not yet confirmed.
Conclusions: There is a high prevalence of left and/or right ventricular dilatation in patients with BAV which is not proportional to the degree of valve dysfunction and 8% of patients had a definitive diagnosis of DCM. This is highly suggestive of a possible genetic link between BAV and DCM, although further genetic investigations will be necessary to confirm our data.

P-93
Decreased Daily Physical Activity in Adults with Congenital Heart Disease

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Objectives: Regular physical activity is known to sustain physical and mental health and reduce all-cause mortality. Despite recent guidelines in the fear of heart failure and sudden death, adults with congenital heart disease (ACHD) were not encouraged to be physically active in the past. By comparing ACHD with healthy controls, this study aimed to examine physical activity among these groups.
Methods: In 100 ACHD (42.7 ± 8.6 years, 50.0% female) and 57 controls (39.1 ± 14.7 years, 68.4% female) daily steps and active minutes (>3MET) were recorded using an activity tracker (Garmin vivofit 3) for one week between September 2017 and November 2018. Comparison was performed using a general linear model corrected for age, sex and body-mass-index.
Results: ACHD were significantly less active per day than controls (ACHD: 8.1 ± 18.5 minutes, controls: 19.5 ± 18.9 minutes, p=.001). Daily steps tended to be reduced in ACHD (ACHD: 9324 ± 3628 steps, controls: 10415 ± 3706 steps, p=.087). ACHD were on average significantly more active on weekends compared to weekdays (weekend: 11.4 ± 19.1 minutes, week: 7.1 ± 9.6 minutes, p<.001). According to the activity tracker's data, 14.8% of the ACHD and 30.4% of the healthy reference cohort reached the WHO guideline of 150 minutes of moderate and vigorous activity a week.
Conclusions: ACHD are significantly less physically active compared to healthy controls, especially regarding intensity. In a medical context most patients have no restriction of physical activity and more advice concerning physical activity can be recommended.

P-94
Serum immunoglobulin G level as an important marker of protein-losing enteropathy in patients with a Fontan circulation

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Objectives: Hypogammaglobulinemia (HG) and lymphopenia (LP) have been observed in Fontan patients. However, data on these complications are scarce. The aims of the present study were to identify risk factors associated with HG and LP in a Fontan cohort.
Methods: Institutional databases were searched to identify Fontan patients in whom serum levels of immunoglobulin G (IgG) and absolute lymphocyte count (ALC) were measured. Lymphopenia was defined as < 1500 cells/uL. Bacterial infectious history of the last 2-years was obtained. PLE remission was defined as a normalisation of serum albumin and fecal alfa-1 antitrypsin (A1AT) levels without having clinical signs and symptoms of PLE. PLE patients not fulfilling these criteria were in active disease.
Results: Fifty-five Fontan patients (37% female, age: 10.7 ± 4.5 years; PLE: n = 14, 25.4%) were included in this study. HG was found in 11/55 patients (20.0%). All of these patients had a diagnosis of PLE and had active disease. Three PLE patients were in remission and had (low) normal IgG levels. In addition, LP was present in 21/55 patients (38.2%). LP was observed in all PLE patients with active disease and HG. LP was not found in PLE patients who were in remission. Six patients (PLE n = 5) suffered from a bacterial infection (pneumonia n = 5; sepsis n = 1). Moderate correlations were found between IgG, albumin levels (R = 0.647, P < 0.0001), and ALC (R = 0.527, P < 0.0001). Moderate inverse correlations were observed between IgG levels, A1AT (R -0.442, P 0.013) and markers of systemic inflammation (R -0.600, P <0.0001). A multiple regression analysis was run to predict serum IgG. Serum albumin, ALC, and WBC significantly predicted IgG levels (F= 21.9, P < 0.0001, R square = 0.569). Fecal A1AT, NTproBNP level and time since Fontan could not predict IgG concentration.
Conclusions: Immune alterations such as HG and LP are common in PLE and non-PLE Fontan patients, respectively. Serum IgG levels can be predicted from albumin levels, ALC and WBC and seem negatively related to active disease in PLE patients.

P-95
Use of clinical, surrogate, and intermediate endpoints in randomized controlled Fontan trials

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Objectives: Randomized controlled trials (RCT) are pivotal for directing evidence-based clinical care. Selection of meaningful endpoints is an essential part of the study design of an RCT. However, no study has systematically examined endpoint selection in Fontan RCTs. In the present study we sought to examine trends in endpoint selection in contemporary Fontan RCTs.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Methods: A search of the PubMed database was conducted using the keywords 'Fontan circulation', Fontan' AND 'randomized controlled trial' OR 'randomized prospective study' to identify all Fontan RCTs published from 2002 to 2017. The following data were extracted from each identified trial: (1) journal, (2) year of publication, (3) study design, (4) intervention, (5) number of patients, (6) number of participating sites and countries, (7) endpoints (primary and secondary), (8) whether the trial met its intended endpoints, and (9) funding sources. Endpoints were categorized as clinical, intermediate or surrogate.
Results: Twenty-two RCT's were found eligible for inclusion. A total of 979 Fontan patients were included in the final analysis. Forty-six primary endpoints were identified. A median of 1 primary endpoint (range 1 - 9) was used per RCT. Eight (17.4%) endpoints were clinical. The majority of these endpoints were categorized as intermediate (n = 25, 54.3%). Change in peak VO2 was most commonly used (n=8). A total of 100 secondary endpoints were identified, mainly categorized as intermediate (n = 47, 47.0%). Change in heart rate (n = 4) was the most frequently used secondary endpoint. Only nine trials (40.9%) met their intended endpoints. Of the 7 RCT's using clinical endpoints, none were able to reject the null hypothesis. Surrogate and intermediate endpoints were frequently combined in the RCTs (n = 15, 68.2%).
Conclusions: This study is the first to demonstrate the heterogeneity and the frequent use of intermediate and surrogate endpoints in contemporary Fontan RCTs. There is a great need to develop validated and standardized endpoints in Fontan research.

P-96

Lifestyles and determinants of perceived health status in Grown-up congenital heart (GUCH) diseases – Results from a national survey in Italy

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Introduction: The rate of grown-up patients with congenital heart disease (GUCH) has been growing steadily among the population, supported by the innovation in both cardiac surgery and medical treatments. GUCH peculiar clinical problems require tailored treatments and care, but a number of analysis about their modifiable risk factors, such as the lifestyles.
However, the information on the epidemiological and clinical aspects of GUCH are poorly described and contradictory. Especially, the lifestyles and determinants of perceived health status among GUCH remains low described worldwide, despite their possible influence on the overall adherence to the follow-up sand consequently clinical outcomes. Accordingly, this study aim to provide the first Italian epidemiological description of the GUCH population lifestyles, identifying the determinants of a poor perceived health status.
Methods: A cross-sectional survey was conducted in an Italian GUCH centre and supported by the Italian Association of GUCH (AICCA). An online survey collected the main socio-demographic and clinical characteristics of the enrolled GUCH, their lifestyles and the perceived health status. The Ethics Committee of our centre approved the study (36/INT/2015).
Results: A sample of 626 GUCH was enrolled (53.9% female; mean age: 35.69±13.49 years; mean BMI: 23.18±4.07 kg/m2). Considering the lifestyles, 65 patients consume tobacco (10.4%), and 81 patients (12.9%) occasionally consume drugs. Roughly, half of the patients (52.1%) perform regular physical activities. The sample reported an adequate received sex education (76.1%) and 40.5% of them use contraceptive methods. Roughly, one patient out every five (18.1%) declared to be low adherent to the ongoing medical treatment. Overall, 53.6% (n=337) and 63.8% (n=401) of the sample have an adequate scores of physical and mental health, respectively. The odds of inadequate perception of physical health increased by more than two times in patients under antiarrhythmic therapies, more than 1.5 times in patients under anticoagulants, and approximately 1.7 times in patients treated with antiplatelets. The odds of inadequate perception of mental health increased by 1.7 time for every year of aging.
Conclusion: This is the first description of the GUCH's lifestyles, underlining illicit use of drugs and some psychological needs of these patients. Further research is necessary.

P-97

Outcomes and quality of life after Ross reintervention: would you make the same choice again?

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Introduction: The Ross procedure was introduced as a long term if not definitive solution for aortic pathology. However, the percentage of reoperation after Ross procedure is not negligible and it can be complex.
Methods: In our single center prospective study, we evaluate the outcomes and the perceived quality of life in patients re-operated after Ross procedure. In addition, a subgroup of patients, in whom a Ross procedure was performed despite an aortic ring of adequate size for mechanical valve implantation, were asked if they would make the same choice again.
Results: Between February 2005 and December 2017, N=64 consecutive patients who had undergone a Ross intervention, were referred for reoperation in our center. Median age was 31 years (10- 56 years). The median freedom from reoperation after the Ross procedure was 136 months (5-271 months). SF-36 questionnaire was administered to the patients at least six months after surgery, and the mean follow-up was 77 months (6-164 months). A total of N=96 procedures were performed. The autograft required reoperation in N=49. Twenty-five patients received a surgical procedure on the right outflow tract. There was not in-hospital nor early mortality. Thirteen patients (20%) had complications. There was one death in the long-term follow-up and one endocarditis medically treated. The SF-36 questionnaire detected good physical parameters and high scores in 95% of patients.
Fifty-two patients (81.2%) had preoperative dimensions of the aortic annulus adequate to receive a prosthetic valve instead of a Ross procedure. The mean age in this subgroup of patients was 22 years (10-48 years) and the median freedom from reoperation after Ross procedure was 137 months (28-271 months). When asked if, with hindsight, they would make the same choice, only 31% replied that they would do it again.
Conclusions: Re-operations after Ross procedure are safe, with low mortality and low but not negligible morbidity. The results in the long-term follow-up are good and the quality of life after reoperation remains high. However, it is our duty to thoroughly explain to patients choosing a Ross procedure, what kind of disappointments they could have in the future.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-98

Surgical management of right partial anomalous pulmonary venous connection in adults: a sigle-centre experience

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Introduction: Right-sided partial anomalous pulmonary venous connection (RPAPVC) to superior vena cava (SVC) is a rare congenital cardiovascular anomaly. Surgery remains the strategy of choice. The main goal is to divert the anomalous pulmonary vein (PV) drainage to the left atrium. We sought to review our short and long-term outcomes after surgical repair of RPAPVC to SVC in adults.
Methods: Retrospective analysis of consecutive patients >14 years old with RPAPVC to SVC operated in our Adult Congenital Heart Unit (ACHU) from May 1996 to December 2017. Diagnosis was made with echocardiography in 100% and cardiac nuclear magnetic resonance in 95%. Catheterization was reserved for cases of doubtful operability and/or presence of cardiovascular risk factors. We indicate surgery if QP/QS>1,5 and no data of irreversible pulmonary hypertension. Different surgical techniques were applied based on localization of RPAPVC into SVC.
Results: We identify 51 patients; mean age: 39±17 years, males: 65%, preoperatory arrhythmias: 20%, atrial septal defect: 80%, persistent left SVC: 16%, mean QP/QS: 2,2/±0,6, mean systolic pulmonary artery pressure: 36±10mmHg). Techniques were; atrial patch closure from right atriotomy (33%), intra-caval baffle with venotomy (61%) and Warden procedure (6%). Eight (15%) patients presented postoperative arrhythmias- one needed permanent pacemaker for complete atrioventricular block. No patient required early percutaneous interventionism (PI) or early reoperation. There was no early mortality. Follow-up was completed in all patients (mean: 5,5±4,4 years, maximum: 17,6 years). PV obstruction occurred in one (2%) patient and SVC obstruction in two (4%). One (2%) patient precised late PI and two (4%) underwent late reoperation- one for pericardial cyst removal. One late death happened due to a non-cardiovascular cause. There was an improvement in NYHA class, tricuspid insufficiency and right ventricle function. Currently, there are no patient with residual PV obstruction and just two (4%) patients with residual SVC obstruction.
Conclusions: Surgical repair of RPAPVC to SVC in adults can be accomplished with excellent results in ACHU. Our choice of surgical approach depending on the localization of anomalous PV seems to be correct. Based on our experience we recommend atrial patch closure for lower drainages and intra-caval baffle for the highest ones.

P-99

Comorbidity and long-term outcome in patients with congenital heart block and their siblings exposed to Ro/SSA autoantibodies in utero

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Objective: Congenital heart block (CHB) may develop in fetuses of Ro/SSA autoantibody-positive women. Given the rarity of CHB, information on comorbidity and complications later in life is difficult to systematically collect for large groups of patients. We therefore used nation-wide health care registers to investigate comorbidity and outcomes in patients with CHB and their siblings.
Methods: Data from patients with CHB (n=119) and their siblings (n=128), all born to anti-Ro/SSA-positive mothers, and from matched healthy controls (n=1,190) and their siblings (n=1,071), were retrieved from the Swedish National Patient Register. Analyses were performed by Cox proportional hazard modeling.
Results: Individuals with CHB had a significantly increased risk of cardiovascular comorbidity, with cardiomyopathy and/or heart failure observed in 20 (16.8%) patients versus 3 (0.3%) controls, yielding a hazard ratio (HR) of 70.0 (95% CI 20.8-235.4), and with a HR for cerebral infarction of 39.9 (95% CI 4.5-357.3). Patients with CHB also had a higher risk of infections. Pacemaker treatment was associated with a decreased risk of cerebral infarction but increased risks of cardiomyopathy/heart failure and infection. The risk of systemic connective tissue disorder was also increased in patients with CHB (HR 11.8, 95% CI 4.0-11.8), and both patients with CHB and their siblings had an increased risk to develop any of 15 common autoimmune conditions (HR 5.7, 95% CI 2.83-11.69 and 3.6, 95% CI 1.7-8.0, respectively).
Conclusions: The data indicate an increased risk of several cardiovascular, infectious and autoimmune diseases in patients with CHB, with the latter risk shared by their siblings.

Key words: Congenital heart block, pacemaker, SLE, Sjögren's syndrome.

P-100

Fourth Decennium after the Arterial Switch Operation for Transposition of the Great Arteries

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Introduction: From 1977 onwards, patients in our hospital (Wilhelmina Children's Hospital) with both simple and complex transposition of the great arteries (TGA) were treated with the arterial switch operation (ASO). Consequently, today, we have a relatively large number of patients with over 30 years of follow-up. In this study, we compared mortality and morbidity in this group with outcome of patients operated more recently and studied echocardiographic left ventricular function.
Methods: A single institution retrospective cohort study was performed on patients who had an ASO for either simple or complex TGA. Data were collected from medical records. Patient cohorts were distinguished in an early era (group A; $\geq$ 30 years of follow-up) and a recent era (group B; $<$ 30 years of follow-up). Type and number of reinterventions as well as standardized echocardiographic follow-up were evaluated.
Results: More than 30 years follow-up (median 33.0, IQR 31.9 – 35.7 years) was available in 48 (of 67) surviving patients (group A). Less than 30 years follow-up (median 13.1, IQR 7.7 – 19.6 years) was available in 208 (of 216) surviving patients (group B). Early survival was best in the youngest group B ($P < 0.001$). Late mortality was rare (1.1%) and similar in both groups. Functionally, all but 3 patients were in functional class I. Reinterventions, corrected for follow-up time, were more frequent in group A ($P = 0.043$), with over all, 48 patients (16.3%) requiring 62 reinterventions. The mode of reinterventions has shifted over time, from surgical to mostly catheter-based ($P = 0.004$). Left ventricular ejection

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

fraction in group A was significantly lower than ejection fraction in group B (52.9 ± 4.1% and 56.7 ± 6.3% resepctively; P = 0.010). *Conclusion:* Early survival improved over time. At follow up, mortality is rare and most patients are in functional class I. Reintervention are performed less frequently and are nowadays mostly catheter-based. Left ventricular ejection fraction is significantly decreased in patients in their fourth decade compared to patients operated on more recently.

P-101
CMR-derived Cardiac Index (CI) is associated with adverse outcomes in patients with single-ventricle Fontan circulation

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Introduction: Fontan patients are exposed to multi-organ dysfunction (liver malignancy, chronic kidney disease, protein-losing enteropathy and others) and low exercise capacity. They also have reduced heart rate reserve. We hypothesized that a MRI-derived cardiac index may be a good tool for an overall assessment of function of Fontan circulation.
Methods: We calculated and investigated a CMR-CI in 34 adult Fontan patients retrospectively. There was no significant relationship between cardiac index and investigated parameters of organs dysfunction. Over a median follow-up of 4 years, 16 events occurred: 9 arrhythmias, 5 non-elective hospitalizations and 1 death.
Results: Average CMR-CI was 3,58 L/min/m2 (min. 2,1 L/min/m2, max. 6,1 L/min/m2). Maximal oxygen uptake ranged from 13,16 ml/min/kg to 28,37 ml/min/kg/, mean 19,14 ml/min/kg. Statistical analysis of correlation between CMR-CI and organ dysfunction parameters was performed. Achieved results are depicted in the table below.

Laboratory tests	N	R	p-value
CI & ALT	34	-0,048402	0,785751
CI & AST	34	-0,085811	0,629426
CI & bilirubin	33	0,080589	0,655726
CI & serum iron level	30	-0,040716	0,830839
CI & creatinine level	34	-0,017049	0,923759
CI & TSH	33	-0,163490	0,363294
CI & Hgb	34	-0,150134	0,396718
CI & Mcv	34	-0,135117	0,446124
CI & RBC Count	34	-0,146101	0,409667
CI & Platelet Count	34	0,239493	0,172509
CI & WBC Count	34	-0,032431	0,855522
Exercie capacity			
CI &pVO2	33	0,272750	0,124612

CI	GFR < 90 ml/min/kg			GFR > 90 ml/min/kg			p
	Mean	SD	n	Mean	SD	n	
	3,425000	0,938749	10	3,642083	0,977250	24	0,508267
CI	Stable NYHA class			Increase of NYHA class			p
	Mean	SD	n	Mean	SD	n	
	3,632083	0,920255	24	3,010000	0,381838	2	0,193856
CI	pVO2 decrease			pVO2 increase			p
	Mean	SD	n	Mean	SD	n	
	3,916000	0,931574	5	3,609444	0,873394	18	0,576056
CI	Lack of nonelective hospitalizations			Nonelective hospitalizations			p
	Mean	SD	n	Mean	SD	n	
	3,754286	0,879509	21	2,870000	0,653758	5	<0,5000
CI	Lack of arrhythmia			Arrhythmia present			p
	Mean	SD	n	Mean	SD	n	
	3,903158	1,041778	19	3,194444	0,692209	9	0,115406

Conclusions: Reduced CMR-CI is predictor of nonelective hospitalizations in this group of patients. CMR-CI does not correlate with organ dysfunction parameters. It suggests development of some degree of multiorgan functional tolerance despite chronic organ hypoperfusion. Whether it results from decreased cardiac output rather than venous congestion should be elucidated in further studies.

P-102
Predictors of left ventricular outflow tract obstruction after conventional repair for patients with interrupted aortic arch or coarctation of the aorta, combined with ventricular septal defect – single-centre experience

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Introduction: Left ventricular outflow tract obstruction (LVOTO) is an important factor affecting survival and reoperation rates after surgical treatment of patients with interruption of the aortic arch or coarctation of the aorta (IAA/CoA) with ventricular septal defect (VSD). The aim of the study was to determine predictors of LVOTO after the repair of IAA/CoA with VSD and to evaluate the relationship between aortic valve (AoV) morphology and the re-intervention rate.
Methods: We conducted a retrospective study of patients after conventional repair for IAA/CoA with VSD at our institution between 1996 and 2017. The pre- and post-operative echocardiographic parameters and re-interventions (surgical and transcatheter) were reviewed.
Results: A total of 52 patients were included in the study [one-stage repair, 47 (90.4%); staged repair, 5 (9.6%)]. The median age at the surgery was 21 days (range 14-27). In the median follow up of 8.31 years (range 6.15-10.27) 8 patients (15.4%) presented with a significant LVOTO; seven of them required reoperation after median period of 2.1 years (range 0.83-8.14). Multivariable logistic regression identified AoV diameter and z-score (OR 0.57, p= 0,035) as predictors of LVOTO.
The mean AoV z-score before the primary repair was significantly smaller in those with LVOTO as compared to those with unobstructed flow from the LV (-3.46 ± 1.56 vs. -1.53 ± 1.57; p=0,0023). At 1-year follow-up, both groups showed an increase in the AoV z-score (-0.78 ± 0.42 and 0,17 ± 0.26), although the difference was not statistically significant (p=0,91). The re-intervention rate, either for LVOTO or reCoA, was higher in patients with AoV z-score ≤ -3 (58.3% vs. 30.8%; p=0.16). Similarly, there was an increased incidence of LVOTO and reCoA in patients, whose aortic annulus was less or equal than patient's weight [kg] + 1.5 mm as compared to those with larger aortic annulus (71.4% vs. 33.3%; p=0,13) [Hirata et al.].
Conclusions: In patients after surgical treatment of IAA/CoA with VSD, the AoV diameter and z-score at the diagnosis are significant risk factors for LVOTO.
With age, AoV growth and z-score improvement is expected. Our data show that small AoV increases the re-intervention rate for reCoA.

P-103
Risk factors for ventricular arrhythmias in tetralogy of Fallot using cardiovascular magnetic resonance

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

CMR data

	Outcome (n =19)	No outcome (n = 415)	P value
RVEDVI	139 ± 47 (102 -171)	107 ± 34 (85 – 121)	0.001
RVESVI	76 ± 35 (55 – 90)	52 ± 23 (38 – 61)	0.0002
RVEF	46 ± 10 (39 – 54)	53 ± 9 (47 – 59)	0.007
PR	24 ± 17 (6-39)	21 ± 17 (4 – 34)	0.480

Mean±SD (interquartile range)

Relative risk for VA

	RR (95% CI)	P value
RVEDVI ≥150ml/m2	5.6 (2.33-13.41)	0.0009
RVEDVI ≥130ml/m2	3.8 (1.57-8.93)	0.004
RVESVI ≥80 ml/m2	5.3 (2.21-12.76)	0.0012
RVEF <40%	0.2 (0.07-0.50)	0.005
PR >40%	1.8 (0.61 – 5.34)	0.290

Introduction: Tetralogy of Fallot (TOF) is the most common congenital cyanotic heart disease. The development of ventricular arrhythmias (VA) and sudden cardiac death (SCD) is the most common cause of late mortality.
Although previously pulmonary regurgitation (PR) was found to be the most common hemodynamic abnormality associated with VA, later cardiovascular magnetic resonance (CMR) based studies did not find this association.
The aim of this study is to investigate the risk factors for VA using CMR in TOF.
Methods: Electronic records of TOF patients and their CMR studies between July 2006 and October 2018 in one center were retrospectively reviewed. Demographic, clinical and CMR data of patients were collected. Outcome data included sustained ventricular tachycardia (VT), SCD as well as aborted SCD.
Results: The total number of TOF patients with complete CMR studies identified was 434. 19 patients developed a positive outcome (14 VT, 3 SCD, 2 aborted SCD) at a median age of 25 (19 – 39) years. Among CMR parameters, the relative risk (RR) for VA was most significant for right ventricular end-diastolic indexed volume (RVEDVI) and right ventricular end-systolic indexed volume (RVESVI) compared to pulmonary regurgitant fraction (PR %) and left ventricular parameters. The highest area under the ROC curve for predicting a positive outcome was 0.721 for RVEDVI,0.755 for RVESVI compared to 0.524 for PR%.
Conclusions: Right ventricular dimensions are the most significant factors associated with the development of VA in TOF patients. PR severity in isolation has no significant influence on the risk of development of VA.

P-104
Coronary computed tomography angiography and echocardiography in children with homozygous familial hypercholesterolemia

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Introduction: Homozygous familial hypercholesterolemia (hoFH) is a rare genetic disease, hallmarked by a lifelong exposure to very high levels of low-density lipoprotein cholesterol (LDL-C). Untreated, patients can experience their first cardiovascular event in the first decade of life. Lipoprotein apheresis (LA) may delay this process, but the required intensity of this invasive therapy is under debate. Early detection and monitoring of subclinical atherosclerosis in these patients is therefore extremely important. We compared the diagnostic yield of low dose coronary computed tomography angiography (cCTA) compared to echocardiography in assessing subclinical atherosclerosis.
Methods: For this single-center cross-sectional study, we included all pediatric hoFH patients that were treated with LA in Amsterdam UMC. All had undergone both echocardiography and cCTA.
Results: Six hoFH patients were included. Median ages at diagnosis, onset of LA and cardiovascular assess-ment (cCTA and echocardiography) were 2.6, 6.5, 10.8 and 11.1, respectively. Echocardiography revealed no signs of atherosclerosis in any of the six patients. In two patients, mild dilatation of the cardiac chambers was detected and two patients showed signs of mitral or aortic insufficiency. On cCTA, however, non-calcified plaques without stenosis were detected in four patients. In two patients calcified coronary plaques were found at the ostia of the right coronary artery (RCA) or the left main coronary artery (LMCA). Aortic root calcifications were found in two patients
Conclusion: In hoFH children, low dose cCTA is superior to echocardiography for the detection of subclinical coronary and aortic root atherosclerosis and should therefore be part of the cardiovascular monitoring in these children, on top of routine echocardiography.

P-105
Echocardiographic screening for rheumatic heart disease in Turkish school children

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Introduction: Although Turkey is one of the country rheumatic heart disease (RHD) is still prevalent, echocardiographic screening for the prevalence of the RHD was not performed before.
The aim of this study was to investigate the prevalence of subclinical RHD in school children (aged 5-18) by portable echocardiograhy in Ankara.
Methods: Auscultation and screening with portable echocardiography was performed by pediatric cardiologist in total 2550 healty students (1339 females, 1211 males, mean age 11.09 years) in six different schools. Echocardiographic studies performed by (Vivid Q N) and they were assessed according to 2012 World Heart Federation criteria for RHD. After reviewing the echocardiographic images, seventy three students were re-evaluated by an advanced echocardiography device (Philips İE33) by two pediatric cardiologists in the hospital.
Results: Evidence of definite subclinical RHD was found in 39 students (15/1000) and of borderline RKH in 20 students (8/1000). No children had clinical symptoms. Only 2 students with RHD had a murmur at cardiac auscultation.The mean age of children diagnosed with RHD and borderline RHD is 12.4 years and 7.5 years, respectively. The risk of RHD was found to be increased 7-fold in girls and children aged between 14-18 years.
Conclusion: This is the first largest single-center echocardiographic screening study for subclinical RHD in Turkish school children. We found frequency of RHD as 15/1000. This finding is similar with the findings of echocardiographic screening studies performed in middle and high-risk populations. We conclude that to decrease the burden of RHD, echocardiographic screening

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

studies are necessary, and longitudinal follow-up of children with echocardiographically diagnosed subclinical RHD is needed.

P-107
Comparison of Aortic Elastic Properties in Stent Implanted and Operated Patients with Aortic Coarctation

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Introduction: The aim of this study was to compare effects of stenting and operation on the aortic stiffness.
Method: Echocardiographic study was used on patients treated with intravascular stenting (n=10) and operated for the coarctation of aorta (n=10). All operations performed by end-to-end anastomosis method without prostetic material. Patients in both groups had no recoarctation. We analyzed left ventricular wall thickness, systolic functions and stiffness index, distansibility index, strain of descending aorta and ascending aorta.
Results: Age range was 10-35 (mean18.4) years in operated group and 10-39 (mean 18.8) years in stenting group. Age at time of stent implantation was 6-36 (mean15.2) years and age at time of operation was 1-25 (mean 9) years. Evaluation time since stent implantation was 3-7 (mean4.6) years and since operation was 3-16 (mean 10.5) years. There was no significant difference in heart rate, systolic blood pressure, systolic functions of left ventricle and aortic valve gradient between the groups. There was no significant difference at diastole and systole left ventricle diameter, interventricular septum diameter, posterior wall thicknes in between groups. Compared with operated group; patients with stent implanted aort coarctation had smaller post stent descending aorta systolic diameters (stenting group: 1.3±0.2 cm; operated group: 1.5 ± 0.4 cm; $p<0.05$) but similar ascending aorta systolic and diastolic diameters. Aortic distensibility of stenting group was found decreased in the proximal ascending aorta (stenting group:4.6± 1.5.10-6cm2/dyne; operated group: 8.2 ± 3.9.10-6cm2/dyne; $p<0.05$) and post stent descending aorta (stenting group: 4.9±3.4.10-6cm2/dyne; operated group: 9.2 ± 3.6.10-6cm2/dyne; $p<0.05$). Aortic strain of stent implanted patients was found decreased in the proximal ascending aorta (stenting group: %16.1± 7.8; operated group: 22.8± 7.1; $p<0.05$) and poststent descending aorta (stenting group: %14.8 ±7.4; operated group: 22.8± 7.1; $p<0.05$). Compared with operated group stiffness index of post stent descending aorta (stenting group: 6.5±3.2; operated group: 2.7 ± 0.9; $p<0.05$) was found increased.
Conclusion: Children who undervent stent implantation have decreased aortic elasticity compared with operated aort coarctation group. This situation may lead to more permanent hypertension or more diastolic dysfunction in stent implanted patients compared with operated aortic coarctation which may develop in the future.

P-108
Athlete heart in children population: echocardiografic findings

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Measurement	Mean Z-score	Z-score >2
IVSd	1,18	20,2%
LVPWd	0,06	3%
LVEDd	0,26	3%
LAD	0,51	9,8%
RVd	0,63	14,3%

Introduction and objectives: Adult athlete heart is defined as the changes developed by routine, systematic and sustained sports training.
We aimed to describe children's athlete heart and to analyze these changes in young hearts through echocardiography.
Methods: 331 children (6 to <18yo, mean 11,92yo, 74% male) under federated sports (football, swimming, tennis, athletism, basketball) were recruited. They performed at least 3hr training/week during more than 1 year long.
Left ventricle (LV) and right ventricle (RV) diastolic diameters and wall thickness were recorded by echocardiography (M-mode) plus left atrium diameter (LA) in apical view. All measurements were indexed by body surface area and analyzed according to type of sport, training time/week and sport practicing exposure.
Results: 118 (33,62%) were football players, 99 (28,21%) swimmers, 58 children (16,52%) were federated in tennis, 40 (11,4%) athletes and 16 (4,56%) basketball players.
Mean training time/week was 7,2h, higher for swimmers and tennis players (>8h/week).
Mean LV interventricular septum Z-score (IVSd) was 1,18. For 20,2% of children, IVSd mean Z-score was >2, particularly those with a higher training rate.
Mean LV posterior wall thickness Z-score (LVPWd) was 0,06. 3% of children had LVPWd Z-score >2, specially the elder ones (14,2 vs 11,9 yo).
Mean LV diastolic diameter Z-score (LVEDd) was 0,26. 3% had LVEDd Z-score >2.
Mean left atrium diameter Z-score (LAD) was 0,51.
Mean right ventricle diastolic diameter Z-score (RVd) was 0,63, increased up to 0,92 in swimmers.
Conclusions: We describe cardiac remodeling in young athletes, more relevant in LV interventricular septum and RV diameter. These changes are probably in relationship with the type of sport and training time.
We suggest follow-up of these young athletes by professionals.

P-109
Functional echocardiographic assessment in congenital diaphragmatic hernia newborns: not only a right ventricle matter

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Objectives: To characterize patterns of preoperative and postoperative ventricular function in infants with congenital diaphragmatic hernia (CDH).
Methods: Infants treated for CDH at a tertiary center during 2017-2018 were included in this prospective, observational study. Echocardiography was performed in the first 12 hours of life, 7 days following surgery, and 30 days of life. Parameters analyzed included measurements of aortic VTI and cardiac output (CO) and

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Values: Median(IQR)	Control(n=16)	CDH 12h (n=5)	p-value*	CDH 1month (n=7)	p-value**
Aortic VTI	11 (10, 12)	7.7 (5.8, 8.2)	**0.004**	14 (11, 14)	**0.011**
Cardiac output	0.6 (0.4, 0.6)	0.3 (0.1, 0.5)	0.064	0.7 (0.6, 0.8)	**0.006**
FAC	55 (48, 63)	47 (35, 48)	0.050	33 (28, 42)	0.230
TAPSE	9 (7, 10)	5.5 (6.5, 7)	0.079	8 (6, 10)	**0.006**
PW E Tric	53 (42, 57)	38 (32, 43)	**0.029**	64 (50, 73)	0.054
PW A Tric	64 (49, 68)	46 (45, 49)	0.050	65 (56, 77)	**0.006**
ED RV area	2.7 (2.5, 3.0)	1.7 (1.4, 2.5)	**0.039**	3.2 (2.7, 3.6)	0.073
ES RV area	1.6 (1.3, 2.0)	1.1 (0.8, 1.5)	0.099	2.1 (1.5, 2.5)	**0.037**
LVEF	66 (63, 71)	75 (70, 83)	0.211	75 (68, 78)	0.667
Septal S'	4 (4, 5)	3.5 (3.3, 3.8)	0.148	4.2 (4.0, 5.4)	**0.029**
MAPSE	5.5 (5, 6)	3.3 (2.5, 4.8)	**0.046**	4.5 (3.5, 6)	0.164
SAPSE	5 (5, 6.8)	3 (2.5, 3)	**0.01**	5 (4, 5)	**0.006**
Ejection Time	195 (185, 210)	156 (150, 171)	**0.009**	180 (156, 185)	0.230
PW E Transmitral	59 (49, 63)	46 (41, 48)	0.064	82 (72, 89)	**0.014**
PW A Transmitral	52 (48, 61)	47 (31, 55)	0.290	79 (72, 105)	**0.011**
Septal E'	5 (4,6)	3.7 (3.4, 4.4)	0.099	4.7 (4.3, 6.2)	0.230
Septal A'	6 (5, 7)	5.7 (4.3, 6.7)	0.617	8.1 (6.5, 8.4)	**0.042**
E/E'	9.5 (6.9, 12)	8.2 (8, 9.4)	0.712	14 (9, 16)	**0.048**
ED LV Volume	4 (3, 5)	1.7 (1.6, 2.5)	**<0.001**	4.5 (3.2, 4.8)	**0.005**
ES LV Volume	1.5 (1, 2)	1.1 (0.7, 1.3)	0.208	1.9 (1.8, 3.0)	**0.048**
Strain Apical Septal	-	-28 (-31, -23)	-	-37 (-39, -36)	**0.036**
Strain Apex	-	-27 (-30, -24)	-	-34 (-34, -34)	**0.036**
Strain ApicalLateral	-	-27 (-28, -25)	-	-32 (-32, -30)	**0.036**

measurements of right ventricle (RV) and left ventricle (LV) systolic and diastolic function, including LV myocardial strain (LVMS). Data obtained at 12h of life were compared with a control group of healthy newborns. Data are presented as median and interquartile range (IQR).

Results: Eight patients were included;7/8 survived to hospital discharge.Lung-to-heart ratio was 52.5% (38, 55). Half required ECMO with a duration of 8.5 days (7.5, 9.5) and an ICU stay of 61 days (38, 82).At birth, CHD newborns had worse CO compared to controls, with both RV and LV systolic and diastolic dysfunction(Table1).After surgery, CDH newborns had significant improvements in aortic VTI and CO; RV systolic function showed significant TAPSE and diastolic function improvement; and LV systolic function increased, including overall volume augmentation.Though trans-mitral flow improved, overall LV diastolic function worsened(Table1).LVMS analysis showed improvement after surgery, specifically in the apical segments, but did not show any significant changes in mid-ventricular GCS.

Conclusions: CDH patients showed reduced cardiac output and global RV dysfunction at birth compared with healthy controls, with clear improvement in overall RV function after surgery. Patients with CHD have abnormal LV diastolic function 1 month after surgery. The abnormally low LV volumes at birth might conceal underlying diastolic dysfunction, which is then unmasked post-surgery.

P-110
Cardiopulmonary Exercise Test Results According to Severity of Structural Heart Disease in Pediatric Patients

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Introduction: Limitation in exercise capacity is present in patients with structural heart disease (SHD) even in the pediatric age group. The best indicator of cardiovascular fitness is peak oxygen uptake (peakVO2) but when it comes to identifying the severity of SHD and prognosis, decreased ventilatory efficiency (nadir of ventilation per unit of carbon dioxide production VE/VCO2) may be a more important variable.

Goals: Our aim was to investigate whether peakVO2 or VE/VCO2nadir on the cardiopulmonary exercise testing (CPET) better reflected the severity of impairment in pediatric patients (6-17 years) with SHD.

Methods: We utilized data from patients who underwent CPET at Mayo Clinic Stress Center between 2011-2016. We classified patients according to SHD severity from 0-3 based on 2018 AHA/ACC Guideline. Differences in peakVO2 and VE/VCO2nadir were analyzed according to SHD linear model (PROC GLM) age, sex, and heart rate (HR) lowering treatment.

Results: As expected, compared to the 343 healthy individuals (185 male, 13.0±3.0yrs) the 169 SHD patients (105 male, 14.6±2.6yrs) exhibited poorer values on several CPET parameters (peakVO2, VE/VCO2nadir, peak HR, HR recovery, HR reserve and peak ventilation); the difference becoming more significant with age and SHD severity. Compared to controls (39.1±10.2mL/kg/min), peakVO2 was significantly ($p<.0001$) lower in all of the SHD groups: 1) 32.0±10.3mL/kg/min; 2) 33.0±8.9mL/kg/min; 3) 31.3±8mL/kg/min, but differences among the SHD groups were not significant. VE/VCO2nadir was lower in the healthy population (26.4±3.1, $p<.0001$) vs. the SHD groups: 1) 27.8±5.0; 2) 27.5±3.9; 3) 30.6±4.8. VE/VCO2nadir for SHD group 3 was signficantly higher than groups 1 and 2 ($p<.0001$).

Conclusion: CPET in patients with SHD provides helpful parameters that better define the clinical stage. The reduced exercise tolerance of SHD patients is defined by a lower peakVO2 and less ventilatory efficiency measured by VE/VCO2nadir. With increasing severity of SHD there is a worsening of ventilatory efficiency though the differences in peakVO2 among SHD groups was not significant. Mechanism of elevated VE/VCOnadir are likely different in pediatric versus adult patients, more related to right-to-left shunting than heart failure.

Keywords: cardiopulmonary exercise testing, pediatric patients, structural heart disease.

P-111
Imaging and laboratory markers associated with quality of life in Fontan patients

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Introduction: With the increase in life expectancy of Fontan patients quality of life has become a fundamental aspect of these patients' follow-up. Recent studies have addressed quality of life in Fontan patients with heterogeneous results. The aim of this study was to assess the importance of variables associated with functional status in quality of life of Fontan patients.

Methods: Fontan patients aged 8 years or older and followed up regularly at our hospital were invited to fill in the Paediatric Quality of Life Inventory 4.0 questionnaire® (PedsQL) at their evaluation at the Fontan Clinics (version according to age). Clinical evaluation, blood tests, transthoracic echocardiogram (TTE) and cardiopulmonary exercise test (CPET) were also performed. The four main scales for quality of life related to health contemplated in PedsQL - physical, emotional, social and academic functioning were evaluated as well as the summary score for psychosocial health.

Results: Fourty-two Fontan patients aged 9 to 36 years of age (median 18.5 years, IQR 12-23) were included. Sixty per cent of patients were male. Completion of Fontan was performed at a median of 6 years (IQR 5-8), and Fontan completion had been performed at a median of 10 years (IQR 7-15) before this evaluation. Physical functioning was reported as worse for female patients (62.42±15.60 vs 73.69±16.17 points, $p=0.0039$). Emotional functioning was positively related to global longitudinal strain (GLS) of the dominant ventricle ($p=0.026$). Social functioning was positively

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

associated with age (p=0.029), with no relation with time since Fontan completion. Academic functioning was positively related to CPET workload (p=0.038). Total quality of life was positively related to age (p=0.032), and negatively related to BNP (p=0.041), with no relation with age since Fontan completion. In a multivariable analysis total quality of life increased 5.14 points for each 10 years of life (p=0.022) and decreased 3.55 points for a BNP increase of 100pg/mL (p=0.028). No variables were related to psychosocial performance.
Conclusions: Variables associated with quality of life in Fontan patients were GLS, BNP and CPET workload. Therapeutic approaches in Fontan patients with an impact on these variables may have a positive influence on quality of life.

P-112
Study of cardiac function and cardiovascular risk factors in pediatric end-stage renal disease

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Introduction: Cardiac disease is a leading cause of morbidity and mortality in children with end-stage renal disease (ESRD). The mortality rate caused by cardiovascular disorders is up to 1000 times higher in children with ESRD compared to healthy children.
Methods: We analyzed a group of 20 pediatric maintenance dialysis patients with a mean age of 14.1 ±3.7 from our tertiary referral center. 10 were on peritoneal dialysis and 10 on hemodialysis, with a mean duration of 30.7 ±26.8 months.
The main focus was the transthoracic ultrasound evaluation of myocardial damage, namely on the presence of left ventricular hypertrophy (LVH). The left ventricular mass (LVM) was calculated and indexed to the power of 2.7 and 2.16, respectively. The type of LVH was assessed using the relative wall thickness (RWT). Statistical analysis was performed using Microsoft Excel 2013. We studied their relationship with traditional and uremia-specific cardiovascular risk factors by calculating the Pearson correlation coefficient and the R2 determination coefficient. The result was considered significant if the corresponding two-tailed p-value was <0.05.
Results: 70% of the patients had LVH, with no significant differences between the two types of renal replacement therapies, 40% presented with eccentric hypertrophy and 30% had diastolic dysfunction. In about 20% of them, the assessment of the ejection fraction and of the shortening fraction showed a reduction <50% and <27%, respectively. There was a significant correlation between the LVM Index (LMVI) and both high Systolic and Diastolic Blood Pressures, with values >95th percentile in 9 patients (45%), despite receiving prior medication. There was a significant negative correlation between the hemoglobin level and the LVMI. Statistically significant correlations between LVMI and elevated phosphorus levels were also obtained, but not for serum lipids, transferrin or homocysteine.
Conclusions: Echocardiography is an extremely useful tool that provides valuable information for diagnosing and monitoring heart disease in patients with ESRD. In our small-sized study group, there was a high prevalence of LVH. Until the possibility of kidney transplantation, which can greatly improve the cardiac function, the management of risk factors (arterial hypertension, anemia, etc.) is required for reducing cardiovascular complications in ESRD pediatric patients.

P-113
Cardiac results of enzyme replacement therapy in Romanian children with mucopolysaccharidosis type I and II

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Introduction: The prevalence and severity of cardiovascular disease in patients with mucopolysaccharidosis (MPS) is high and progressive, consisting in severe cardiac valve disease and ventricular hypertrophy. Enzyme replacement therapy in MPS may improve the organ impairment. The aim of the study was to characterize the cardiac disease and the results of enzyme replacement therapy in children with MPS type I and MPS type II.
Methods: We evaluated 23 patients with mucopolysaccharidosis (MPS): 5 patients with MPS type I and 18 patients with MPS type II. We assessed the function of valves, left ventricular chamber dimensions, septal and posterior ventricular wall thicknesses, systolic and diastolic function of the ventricles, pulmonary hypertension at every 6 months. The treatment of these patients consisted in weekly administration of recombinant form of human alpha-L-iduronidase in dose of 0.58mg/kg iv weekly for 5 patients with MPS type I and iduronate 2-sulfatase in dose of 0.5mg/kg, iv weekly for 16 patients with MPS type II.
Results: At diagnosis, all patients presented echocardiographic alterations. Mitral valve thickening with variable grades of regurgitation was diagnosed in all patients, aortic regurgitation was present in 56% of patients and mitral stenosis in 9% of patients. Left ventricular hypertrophy was diagnosed in 35% of patients. Mild pulmonary hypertension was present in 17% of patients. The mean age of the patients at starting therapy was 6.1 year. The treatment results after 2 years were: valvular heart disease were stable in 67% of patients, mild improvement was present in 19% of patients and aggravation in 14% of patients. Ventricular hypertrophy remained unmodified in 67% of patients and worsened in 11% of patients. After 4 years of treatment the results were: valvular heart disease were stable in 67% of patients, mild improvement was present in 14% of patients and aggravation in 19% of patients. Ventricular hypertrophy remained unmodified in 55% of patients and improvement was recorded in 45% of patients.
Conclusions: The most prevalent cardiac changes in children with MPS are valvular lesions. Enzyme replacement therapy had little effect on cardiac disease in children with MPS.

P-114
Postnatal screening for congenital heart disease: Clinical findings, Pulsoximetry, Echocardiography

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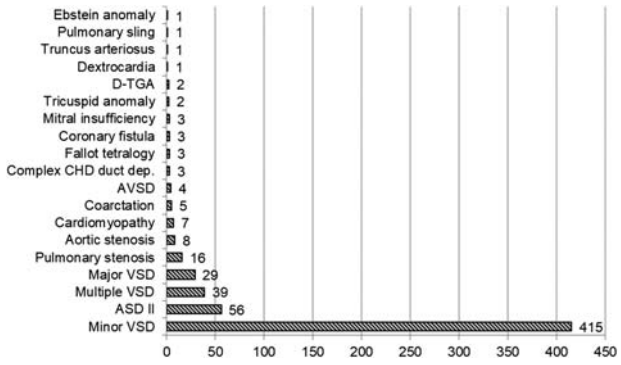
Introduction: Congenital heart defects(CHD) are the most common congenital malformations. Echocardiography performed by a pediatric cardiologist is regarded as the gold standard, detecting even small cardiovascular defects. The primary objective of newborn screening is the pre-symptomatic identification of life-threatening CHD in order to achieve a timely diagnosis before collapse or death occurs.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



Methods: Supported by the government of Hessen, and in written informed consent of the parents, screening echocardiograms (ECHO) were offered for a period of eleven years for all babies who were born between August 1, 2007 and November 30, 2018 at a large maternity hospital managed by obstetricians. The population of neonates comprised those who were unremarkable during pregnancy and who were born at term. The pulse oximetry readings(OXI) were taken between 2nd and 4th hour of life. An experienced pediatric cardiologist performed the echocardiograms using a 10 MHz transducer within the first days (median: 27 hours) of neonatal life after the clinical examination(CLIN). In this study, the results of ECHO were compared with those obtained from CLIN and OXI (Sp02 < 95%).
Results: 13050 neonates(86%) could be included. 599 cases (45,9/1000) of CHD were detected by ECHO: 18 critical, life-threatening CHD(1,4/1000), 155 hemodynamically significant CHD(11,9/1000), 415 minor VSD(31,8/1000) and 11 other defects. Only 16% of the congenital heart defects were detectable by CLIN, 3% by OXI. The sensitivities with regard to the critical CHD were 100%(ECHO), 39%(CLIN), 33%(OXI), 56% (CLIN+OXI). In 16% of the neonates, a fetal echocardiography was performed as a result of the patient history or family history. No congenital heart defect was detected antenatally which was a prerequisite of this maternity hospital.
Conclusions: OXI is an additional tool to detect missed diagnoses of CHD in early life. ECHO is expensive and personnel-intensive, but the only strategy to rule out missed diagnoses of CHD.

P-115
Subclinical early systolic abnormalities are apparant already after low cumulative anthracycline doses in childhood acute lymphatic leukemia

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Objectives: Aim of the study was to assess the acute effect of chemotherapy on cardiac function of children with acute lymphatic leukemia (ALL) short before and after administration of Daunorubicin in the first chemo block. This chemo block starts immediately after diagnosis of ALL and lasts for four weeks.
Methods: 24 children (11 female, 13 male) diagnosed with ALL and treated with AIEOP-BFM ALL 2009 protocol were enrolled in a prospective study. Daunorubicin (DNR, 30 mg/m2/dose in 1 h i.v.) was administered on day 8, 15, 22 and 29 of Block Ia. Echo was performed and NT-pro-BNP and Troponin I was analyzed on day 8, 15, 22 and 29 short before and short after infusion of Daunrorubicin. Transthoracic echocardiography Images were obtained by a GE Vivid 7 and GE E95-scanner with a 4 MHZ and 6 MHZ transducer according to age. Off-line analysis was performed by EchoPAC.
Results: No patient developed heart failure or significant heart problems. No patient developed pathological Ejection fraction-(EF) or fractional shortening (FS) values. EF and FS-values did not decrease over time. In contrast longitudinal strain values showed from day 8 on a decrease over time in absolute values. Already there was a significant decrease in longitudinal strain in the basal septal and mid septal segment over time. NT-pro-BNP-values were in most cases elevated short after chemo but did not correlate with other values. Troponin I-values were not elevated after administration of Daunorubicin.
Conclusion: This is the first study focusing on the cardiac effects of Anthracyclines during the first four weeks of chemo (Block Ia). With a total of 8 echo examinations per patient meanwhile four weeks cardiac function was frequently analyzed. Results showed that abnormalities of systolic function detected by longitudinal strain are already apparent after administration of low cumulative doses of Anthracycline. Strain seems to be more sensitive in comparison to EF and FS. The fact that Troponin I values were not elevated is maybe a sign that myocardial cell apoptosis did not take part yet. The elevated NT-pro-BNP-values short after chemo are most likely an effect of hyperhydration on chemo day.

P-116
Arterial stiffness and myocardial functions in children with isolated bicuspid aortic valve

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Objectives: Due to the deficiency of nitrite oxide synthase, patients with bicuspid aortic valve are at risk for endothelial dysfunction and atherosclerosis as well as valve dysfunction and proximal aortic dilatation. In this study, we aimed to investigate the presence of arterial stiffness and early atherosclerotic changes in children with bicuspid aortic valve.
Methods: Forty five children with isolated BAV and 34 healthy controls with TAV matched by gender, age and body surface area were studied. Patients with aortic valve velocity > 1.7 m/s and more than mild aortic regurgitation (AR) were excluded. Aortic valve morphology, aortic root and ascending aorta dimensions were detected with two-dimensional echocardiography. Left ventricular myocardial functions examined with M-mode and tissue Doppler echocardiography. Bilateral carotis intima media thickness were measured with lineer prob. Aortic bood pressure, augmentation index (AIx) and pulse wave velocity (PWV) measured with ambulatory oscillometric device (Mobil-O-Graph, IEM, Germany).
Results: There were no differences in lipid profile and blood glucose. Echocardiographic parameters were also similar except aortic diameters at four levels (annulus, valsalva sinuses, sinotubular junction, ascending aorta) which were larger in BAV patients (18.4 ± 2.9mm v.s 16.4 ± 2.1 mm, p<0.001, 24.8 ± 4.2 v.s 23.1 ± 2.7, p<0.001, 21.2 ± 4.3 v.s 18.1 ± 1.9, p<0.001, 25.6 ± 5.2 v.s 20.7 ± 2.6, p=0.04, respectively). Compared to controls, the E/Em ratio and tissue Doppler derived myocardial performance index were higher in BAV patients (8.9 ± 2.5 v.s 7.8 ± 1.4, p=0.02, 0.39 ± 0.005 v.s 0.33 ± 0.04, p<0.001, respectively). There were no differences in terms of carotid intima media thickness and central blood pressure values. Although there was a statistically insignificant decrease in terms of pwv, Alx was detected lower in BAV patients.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusions: Although there were findings associated with aortopathy and myocardial dysfunction in patients with well function bicuspid aortic valve even in childhood, no evidence of endothelial dysfunction were detected.

P-117
Transposition of the Great Arteries and Intact Ventricular Septum: postnatal morphological and functional echocardiographic patterns

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Introduction: To improve the understanding of the pathophysiology of Transposition of the Great Arteries with Intact Ventricular Septum (TGA/IVS) and the cardiac remodeling occurring from the fetal life we performed a morphometric and functional echocardiographic assessment in fetuses and newborns. Additionally, we studied the echocardiographic differences in those who developed persistent pulmonary hypertension (PPHT) before the arterial switch operation.
Methods: Prospective case-control study performed in a tertiary referral center, including fetuses and newborns with diagnosis of TGA/IVS during a three year period. Morphometry and systolic and diastolic function parameters were compared with age and body-surface matched healthy controls.TGA/IVS patients were classify in PPHT and non-PPHT groups.
Results: (Table): Twenty one TGA/IVS patients were included during the study period. Morphometric and functional echocardiographic data is shown in the Table. Compared to controls, fetuses with TGA/IVS had bigger right atriums and aortic valves. Both right and left systolic and diastolic function were slightly increased with overall higher aortic cardiac output.
Postnatally, TGA/IVS showed bigger atriums and globulous right ventricles with bigger aortic valves. Heart rate and overall function was significantly increased resulting in both aortic and pulmonary higher cardiac output. In fetal life, patients with PPHT had smaller foramen ovale (5.8 vs 7.8mm) with higher bulging ratios (0.60 vs 0.45) compared to non-PPHT. The pulmonary velocity (84.5 vs 117cm/s), pulmonary index (2.91 vs 4.4 ml/Kg/min) and QP/QS (0.83 vs 0.94) were lower.

Table: Echocardiographics parameters in TGA/IVS in fetal and postnatal life

	Fetal TGA (21)	Controls (50)	*p*	Neo TGA (21)	Controls (63)	*p*
HR (bpm)	140 ± 8	142 ± 10	0.427	149 ± 13.9	126 ± 18	*<0.001*
Aortic CO (ml/min)	662 ± 370	388 ± 192	*0.001*	1146 ± 436	685 ± 225	*<0.001*
Pulm. CO (ml/min)	515 ± 213	454 ± 204	0.223	1061 ± 330	738 ± 195	*<0.001*
QP/QS	0.89 ± 0.36	1.26 ± 0.50	*0.005*	1.1 ± 0.7	0.95 ± 0.3	0.884
RA area (cm2)	2.42 ± 0.76	1.84 ± 0.73	0.022	3.1 ± 1.1	2.1 ± 0.4	*<0.001*
LA area (cm2)	1.74 ± 0.54	1.70 ± 0.61	0.703	2.6 ± 0.9	2.0 ± 0.4	*<0.001*
AV diam (mm)	6.94 ± 1.44	5.56 ± 1.42	*0.001*	9.2 ± 0.9	7.7 ± 0.6	*<0.001*
PV diam (mm)	6.61 ± 1.17	6.11 ± 1.52	0.185	7.9 ± 1	8.1 ± 0.6	0.144
Right FAC (%)	27 ± 12	28 ± 11	0.742	49 ± 7	29 ± 23	*<0.001*
Tricuspid S' (cm/s)	8.8 ± 1.9	7.6 ± 1.3	*0.028*	7.7 ± 1.8	5.6 ± 1.5	*<0.001*
Tricuspid E (cm/s)	46 ± 8	43 ± 8	0.164	88 ± 29	51 ± 15	*<0.001*
Aortic vel (cm/s)	82 ± 18	83 ± 15	0.349	98 ± 25	75 ± 15	*<0.001*
Left EF (%)	71 ± 14	62 ± 11	*0.035*	71.8 ± 8.2	63.7 ± 8.4	*0.001*
Mitral S' (cm/s)	6.8 ± 2.1	6.8 ± 1.2	0.323	6 ± 1.7	4.4 ± 1.1	*<0.001*
Pulm. vel (cm/s)	94 ± 19	70 ± 14	*<0.001*	114 ± 33	81 ± 12	*<0.001*
Mitral E (cm/s)	41 ± 6	38 ± 8	0.170	91 ± 26	58 ± 15	*<0.001*

TGA/IVS: Transposition of the great arteries and intact ventricular septum; CO:cardiac output; RA:right atrium; LA: left atrium; AV:aortic valve; PV:pulmonary valve; FAC:fractional change area; QP/QS calculated as ratio between pulmonary and systemic cardiac output; Data shown as mean ± SD.

Conclusions: TGA/IVS patients show morphometric and functional changes of increased overall volume and output predominantly in the aortic component from fetus to newborn, probably due to compensatory mechanisms secondary to brain hypoxia. Patients with PPHT have lower pulmonary output values already present in the prenatal period. Therefore, measurement of QP/Qs in the prenatal period might help the early recognition of PPHT patients.

P-118
Functional Outcome in Children and Adolescents with Isolated Left-to-Right Shunt

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Introduction: Atrial (ASD) and ventricular septal defects (VSD) represent the two most common congenital heart diseases (CHD) and belong to the simple CHDs. Nevertheless, they represent a chronic diseased population and these patients have increased long-term functional impairments and higher cardiac morbidity.
Objectives: The objective of this study was to investigate several functional outcome measures in children with ASD and VSD in comparison with a healthy control group (CG).
Patients and Methods: From May 2014 to October 2018, we examined 148 patients (72 girls, 11.7 ± 3.6 yeas) with isolated shunts (ASD: 53%, VSD: 47%) for their Health-Related Physical Fitness (HRPF), arterial stiffness, Intima-Media Thickness (IMT) and Health-Related Quality of Life (HRQoL). Native condition was present in 39.7% interventional closure of the defect was performed in 26.5% and surgical closure in 33.7% patients. HRPF was tested by five tasks of the FITNESSGRAM®. The functional arterial stiffness measures, central systolic blood pressure and pulse wave velocity (PWV) were analyzed with an oscillometric device. Structural changes were characterized by IMT of the Arteria carotis communis. HRQoL was assessment of a subjective perspective with the KINDL questionnaire. For comparison, a CG of 2002 children (48.9% girls, 12.8 ± 2.8 years) was recruited within two recent school projects.
Results: After adjustment for age and sex, children with ASD and VSD presented significantly lower HRPF (z-score ASD: -0.49 ± 0.72 p<.001; z-score VSD: -0.69 ± 0.072; p<.001) compared to the CG. Transferred into percentiles, VSD were on the 25th and ASD on the 31th percentile. Structural and arterial stiffness measures did not differ from CG, nor did HRQoL.
Comparing children with ASD and VSD there were no differences at all in-between these two groups. Regarding the surgical history of the shunts (native, interventional closure, surgical closure), there were also no difference in-between the three states.
Conclusion: Children with ASD or VSD have impaired HRPF but fortunately, they have no other functional and structure limitations in terms of arterial stiffness measures and no reduced HRQol. Early childhood sports promotion would be a good intervention to counteract these restrictions in HRPF at an early stage.

P-119
Cardiac magnetic resonance imaging and disease progression in young patients with hypertrophic cardiomyopathy

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Introduction: Cardiac MRI including stress perfusion provides additional information for assessing sudden cardiac death risk in hypertrophic cardiomyopathy patients. However, the pathophysiological meaning of those findings in young patients is not fully inspected.
We investigated the relationship between CMR findings and clinical feature to speculate the mechanism of disease progression.
Method: From September 2011 to September 2016, 17 patients, with a mean age of 14.7 years, with hypertrophic cardiomyopathy underwent comprehensive cardiac MRI (1.5-T Philips Achieva). Cine images, stress and rest perfusion images, and late gadolinium enhancement (LGE) images were acquired successively. Adenosine triphosphate was intravenously injected at a dose of 0.14 mg/kg/min during stress perfusion. Both perfusion images were acquired by infusion of 0.1 mmol/kg meglumine gadopentetate at an infusion rate of 3.5 ml/s respectively.
Result: Stress induced perfusion defect (SPD) was detected in 10/15 patients (67%). LGE was recognized in 14/16 patients (88%). Of those with LGE, the extent was 10±13% of the myocardial mass. Left ventricular mass index was 162±61% of normal value and ejection fraction was 66±10%. In five patients with a history of syncope or ventricular fibrillation, LV mass index and plasma brain natriuretic peptide (BNP) level were significantly higher than in those with no history (206 vs 144% of Normal, p = 0.048, 635 vs 130 pg/ml, p=0.002, respectively). LV mass index was also higher in those with SPD than in those without SPD (184 vs 129% of Normal, p = 0.041), while LGE extent was not significantly different. Out of 3 patients who underwent the second cardiac MRI 2 years after the initial examination, 2 patients with SPD presented the expansion of LGE area along with plasma BNP elevation(LGE extent; 6 to 13%, 4 to 8% and BNP; 107 to 274, 100 to 280 pg/ml, respectively).
Conclusion: Microvascular dysfunction, which is proportional to magnitude of hypertrophy, can be one of the key role players in worsening disease severity. Comprehensive and periodical cardiac MRI including stress perfusion can be feasible and clinically valuable in young patients with hypertrophic cardiomyopathy.

P-120
Left ventricular volumes and function is affected by the cardiac fibrosis in patients with Becker and Duchenne muscular dystrophies in CMR

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Introduction: Duchene Muscular Dystrophy (DMD) and Becker Muscular Dystrophy (BMD) are chromosome X-linked dystrophinopathies affecting skeletal, cardiac and respiratory muscles. Cardiac dysfunction is among leading causes of morbidity and mortality in this group of patients. Despite Cardiovascular Magnetic Resonance (CMR) is considered a useful tool for evaluation of cardiac function and fibrosis, in DMD and BMD patients the data is still scarce.
Methods: Of 79 patients with genetically confirmed diagnosis, 41 (aged 12.0 ± 3.1 years, DMD 88%, n=36, BMD 12%, n=5) were qualified and successfully examined using CMR. Disqualification criteria was age < 6 years, autism or metal implants. CMR protocol included LV dimensions, stroke volume (LVSV), ejection fraction (LVEF) measurement in short axis, and late gadolinium enhancement (LGE; 10–15 minutes after contrast injection) to provide fibrosis assessment. The obtained values were indexed to BSA and normalized (z-score) according to reference data published by Kawel-Boehm. Data is presented as mean ± standard deviation or median (range) dependently on the distribution. Chi-square test, Pearson and Spearman correlations were employed.
Results: Left Ventricle End Diastolic Volume index (LVEDVi) was 63.6 ± 17.4 ml/m2 and was abnormal in 24% (n=10). Left Ventricle End Systolic Volume index (LVESVi) was 30.0 ± 9.0 ml/m2, abnormally high in 12% (n=50) and abnormally low in 2% (n=1). Left Ventricle Mass index (LVMi) was 54.0 ± 12.2 g/m2 and normal in 93% of patients (n=37). LGE was assessed in 39 patients and was positive in 38% (n=15), most often in mid-anterolateral (38%, n=15), basal-anterolateral (36%, n=14), basal-inferolateral (31%, n=12), mid-inferolateral (26%, n=10) and apical-lateral segments (18%, n=7). LVSVi was 37.0 ± 10.8 ml/m2, abnormally low in 39% of cases (n=16), and LVEF was 58% ± 6.4%, low in 44%, n=18. Older patients had significantly lower LVEDVi-z (r=-0.41, p=0.008) and LVSVi-z (r=-0.50, p<0.001 respectively). LGE is significantly more prevalent in older patients (p<0.001). Patients with positive LGE had significantly lower LVSVi-z (p=0.022) and LVEF (p<0.001).
Conclusions: Fibrosis advances with age and DMD/BMD progression, causing worsening of cardiac function by limiting LVEDV and LVSV. The effect of pharmacotherapy is subject of a separate study.

P-121
Severity of myocardial dysfunction in Familial Mediterranean Fever, can it discriminate homozygous from heterozygous cases?

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Introduction: Familial Mediterranean fever is the most common auto-inflammatory disorder. Myocardial dysfunction in the context of FMF might be the result of several factors: persistent inflammation, amyloid deposition, medications involved in control of disease activity.
Methods: 40 patients (Group 1: G1) with FMF and 40 healthy controls (Group 2: G1) have been examined with Tissue Doppler Imaging and Speckle Tracking Echocardiography for examination of Left ventricular Systo-diastolic functions, demographic, clinical and genetic data of cases have been recorded as well. Patients have been furtherly subdivided into Group 1A (G1A) with homozygous mutation and Group 1B (G1B) with heterozygous mutation.
Results: Left ventricular diastolic dysfunction have been depicted in cases with FMF, this dysfunction was more profound in cases with homozygous mutation as evidenced by higher LV E/E' ratio in cases with homozygous than heterozygous FMF mutation (G1A: 15±3 vs.G1B: 6.57±1.2, P<0.001). Moreover subtle systolic LV dysfunction has been found in cases with FMF, LV systolic function was even lower in cases with homozygous mutation as evidenced by a lower LV GLS (global Longitudinal Strain) in such cases (G1A: -11.2±2.2 vs. G1B: -20.2±4.2, P<0.001). LV GLS

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

proved even to be highly predictive for homozygosity in cases with FMF with sensitivity of 94%.
Conclusion: The aforementioned findings point towards the occurrence of systo-diastolic dysfunction early in the course of FMF, cases with homozygous mutation are more at risk, this might be related to more severe inflammation and subsequent amyloid degeneration.

P-122
Cardiac magnetic resonance feature tracking in repaired Fallot patients predicts ventricular tachycardia but not deterioration of ventricular function

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Introduction: Main determinants of morbidity and mortality in patients with repaired Tetralogy of Fallot (TOF) are ventricular tachyarrhythmia's and deterioration of right ventricular (RV) function. Myocardial strain has shown to be predictive of outcome in several cardiovascular diseases, including congenital heart diseases. The aim of our study was to evaluate the predictive value of CMR-derived strain parameters in repaired TOF patients for developing ventricular tachycardia (primary outcome) and deterioration of ventricular function (secondary outcome).
Methods: Patients with repaired TOF who underwent cardiac magnetic resonance (CMR) investigation between January 2007 and March 2016 were included. Strain and strain-rate of both ventricles were assessed using CMR feature tracking. Primary outcome was a composite of the occurrence of sustained ventricular tachycardia (VT) or symptomatic non-sustained VT requiring invasive therapy. Univariate Cox-regression analysis was performed, after which all significant parameters were fit into a multivariate Cox-regression analysis model. Secondary outcome was analyzed in a subgroup of patients that underwent a second CMR after 1.5 to 3.5 years. Deterioration was defined as reduced RV ejection fraction (EF) of ≥10%, reduced left ventricular (LV) EF of ≥10% or increased indexed RV end-diastolic volume (EDVi) of ≥30mL/m2 compared to baseline. Patients marked as 'deteriorated' were 1:2 propensity-score matched at baseline, based on conventional mass/volume CMR measures to patients without ventricular deterioration. Strains and strain-rates were compared between groups to assess whether these parameters precede changes in conventional measures.
Results: 172 patients (median age 24.3 years, 54 patients <18 years) were included. Throughout the median follow-up of 7.4 years, 9 patients (4.5%) experienced the primary endpoint of VT. In the multivariate model, LV systolic circumferential strain-rate was independently predictive of primary outcome with an area under the curve in receiver-operator characteristic (ROC) analysis of 0.79. 70 patients underwent a serial CMR, of whom 14 patients (20%) showed ventricular deterioration. Myocardial strain and strain-rate parameters at baseline did not differ between the 'deteriorated' and 'not-deteriorated' group.
Conclusions: In repaired TOF patients, LV systolic circumferential strain-rate has a strong and independent predictive value of developing VT. No predictive value of strain parameters for deterioration of ventricular function could be demonstrated.

P-123
Determinants and Frequency of Left Ventricular Funktion and Remodelling in Patients with corrected Tetralogy of Fallot

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Objectives: The aim of this study was to identify in asymptomatic Tetralogy of Fallot (ToF) patients after repair the prevalence and determinants of impaired left-sided cardiac function and adverse ventricular remodeling, and the relation of LV dysfunction and remodeling with exercise tolerance.
Methods: In a cross sectional study, 103 ToF patients (median age 16.3 years) in NYHA class I, with surgical repair at a median age of 1.1 years, and 63 age-matched controls were studied. LV, RV function and geometry, LV myocardial extracellular volume (ECV), and left atrial (LA) function were quantified with cardiac magnetic resonance. Peak oxygen consumption (pVO2) was measured by standardized cardio-pulmonary exercise test.
Results: ToF patients had lower LV ejection fraction (EF) ($p=0.001$; 49% below age-adjusted 5th percentile for controls), lower LV mass index ($p=0.003$), lower LV mass-to-volume ratio ($p<0.01$), and impaired LA function. RV mass-to-volume was the best predictor for LV systolic dysfunction and for a lower LV mass-to-volume ratio. LV ECV was higher ($p<0.001$), particularly in females, and associated with subnormal pVO2 ($p=0.037$). A pVO2 below the 3rd percentile reference level was more likely with decreasing LV EF ($p=0.008$), and LV mass-index ($p=0.024$), but independent of RV EF.
Conclusions: In NYHA class I ToF patients, frequent impaired systolic and diastolic LV function, LV remodeling with LV atrophy, a decreased mass-to-volume, and extracellular matrix expansion suggest cardiomyopathic changes. The best predictor for LV systolic dysfunction was the RV mass-to-volume ratio. The subnormal peak oxygen consumption indicates that monitoring of LV status is important for long-term prognosis.

P-125
Assessment of pulmonary valve pulse wave velocity in children and teenagers

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Introduction: Echocardiographic diagnosis in childhood and adolescence has a number of features, primarily due to the need to assess virtually all dimensions in accordance with the weight of the child, as well as the morphogenesis of individual heart structures in different periods of life.
The data on the maximum values of velocity on the pulmonary valve (PV) presented in the literature are often not true. The recorded rates exceed the recommended values even in the absence of pathology of the cardiovascular system, and in the presence of

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

defects of the septum (ASD, VSD) or restless behavior of the child are even greater.
Methods: In order to clarify the situation, we made an attempt to revise the maximum normal speed indicators (Vmax) for PV in childhood. All studies were performed on an ultrasonic device Medison AccuvixV10 by an experienced physician with more than 10 years of experience.
Results: A retrospective analysis of the protocols of 2030 echocardiographic studies was performed, which was performed for children aged 2 weeks to 16 years. Children with ASD, VSD, PDA were not included in the study. The results of the Vmax estimation on the PW-mode, taking into account the body weight of the children surveyed, were as follows:
3,3-9,9 kg - 1,06+0,27 m/s; 10.0-14.9 kg - 1.07+0.25 m/s; 15.0-19.9 kg - 1.02+0.31 m/s; 20.0-24.9 kg - 1.0+0.29 m/s; 25,0-29,0 kg - 1,0+0,26 m/s; 30.0-39.9 kg - 1.0+0.25 m/s; 40.0-60.0 kg - 1.0+0.3 m/s.
The correlation analysis made it possible to reveal only a weak negative connection of Vmax to a PV with age ($r=-0.15$, $p<0.95$), and a moderate positive correlation Vmax on a PV with Vmax on aortal valve ($r=0.43$; $p<0,05$). In the latter case, the explanation may serve as a hyperkinetic type of hemodynamics, more often observed in childhood. In general, for the group, the average values of Vmax for PV were 1.03+0.25 m/s.
Conclusions: The maximum normative speed indicators for PV in childhood should be revised, and set at a minimum within 0.78-1.38 m/s (for M+2δ), and this is only for the PV. For the trunk of LA speeds can be somewhat larger.

P-126
Fontan associated liver disease - prevalence of ultrasound and laboratory abnormalities in different age groups

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Introduction: Liver fibrosis is increasingly recognized as a potentially serious morbidity associated with Fontan circulation (Fontan associated liver disease, FALD). The purpose of this study was to assess the prevalence and progression of liver abnormalities with standard investigations.
Methods: Fontan patients were screened for liver abnormalities by abdominal ultrasound and routine laboratory tests. Patients were divided into three groups based on follow-up since Fontan surgery (<5 years, 5 to 10 years, >10 years). Laboratory test were interpreted based on age and gender adjusted reference values.
Results: 197 Fontan patients seen for routine follow-up between January 2013 and November 2018 who had abdominal ultrasound together with routine laboratory tests were included. Of them, 139 (70.6%) were re-investigated after a median follow-up of 2.4 (IQR 1.4-3.3) years. Median age at latest follow-up was 12.1 (IQR 8.7-16.7) years, the follow-up since Fontan surgery 9.4 (IQR 6.1-14) years. Sonographic signs of fibrotic changes included heterogeneous parenchyma, surface nodularity or hyper-echoic lesions in 103 (52%) patients. The prevalence of fibrotic changes increased with longer follow-up since Fontan completion (<5 years: 9/30 (23%); 5-10 years: 31/65 (48%); >10 years: 63/93 (68%); $p<0.001$). New or progressive fibrosis was seen in 24 of 63 (38%) patients with repeat ultrasound after 2.8 (IQR 1.5-3.2) years. Thrombocytopenia, a feature of portal hypertension, was observed in 3.2% and 2.9% of cases with less than 5 or 5 to 10 years of follow-up, but in 27.4% with more than 10 years of follow-up ($p<0.001$). Elevated Gamma-glutamyl-transpeptidase (gGT) levels were common in all groups, but less frequent with more than 10 years of follow-up (<5 years: 90%; 5-10 years: 97%; >10 years: 72%; $p<0.001$. Mean platelet count was lower (222 ±84/nl vs. 242 ±91/nl, $p=0.041$) and gGT levels were higher in patients with sonographic signs of fibrosis (67 ±42 U/l vs. 53 ±37 U/l, $p=0.002$), while absolute values of other laboratory data showed no differences.
Conclusions: The prevalence of ultrasound and laboratory abnormalities suggestive of FALD increases with time since Fontan surgery. Among laboratory abnormalities the platelet count might serve as a surrogate for progression of liver changes.

P-127
Change in global strain during the first months of life in children born after intra-uterine growth restriction

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Background and objectives: Being born small for gestational age (SGA) may be associated with cardiovascular disease and metabolic disorders in adult life. Underlying changes may be detected early in life. We have assessed left ventricular function in children from suspected intra-uterine growth restriction (IUGR) pregnancies compared to non-affected controls shortly after birth and after 3-4 months.
Subjects and methods: In this prospective controlled study, pregnancies with IUGR > 22% weight deviation were identified by ultrasound examinations due to suspected IUGR. Control pregnancies were identified at routine ultrasound examinations at gestational week 18-20.
Echocardiography (Vivid E9, GE Ultrasound) analysed with Velocity vector imaging (VVI, Siemens Medical Solutions) was used to determine cardiac motion by tracking the grey scale image. Longitudinal velocity, displacement and strain were assessed. Standardized 4-chamber loops were recorded at birth (12-72 hours) and at 3-4 months after delivery for blinded off-line analysis.
Results: We included 20 infants with IUGR [mean (SD) prenatal weight deviation -30.6 (10,48)%, BW 2.0 (0.8) kg, BW SDS -2.6 (1.1), GA 36.3 (3.9) weeks] and 35 non-affected controls [BW 3.4 (0.4) kg, BW SDS 0.00 (0.8), GA 39.8 (1.5) weeks)]. At birth, global strain did not differ between the groups [-16.5 (2.9) % and -16.7 (3.3) %, respectively]. In IUGR, no change in strain at 3-4 months was observed [-17.4 (2.5) %] while controls significantly increased [-18.9 (3.8) %, $P= 0.002$] resulting in a lower global strain in IUGR compared to controls ($P = 0.041$). Longitudinal velocity and displacement were significantly different at birth and at 3-4 months. However, this difference was lost by correction for left ventricular length.
Conclusions: Our data suggest that children with IUGR have less pronounced development of global strain over the first 3-4 months of life resulting in less systolic deformation which could indicate a propensity for cardiovascular disease in adult life. We found that IUGR related differences in myocardial longitudinal velocity or displacement of basal segments, also previously reported, were related to cardiac size. Global strain, purportedly independent of

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

heart size may be a better marker of changes in cardiovascular function in early life.

P-128

Myocardial work analysis in children with Kawasaki disease: an early diagnostic tool of myocardial dysfunction

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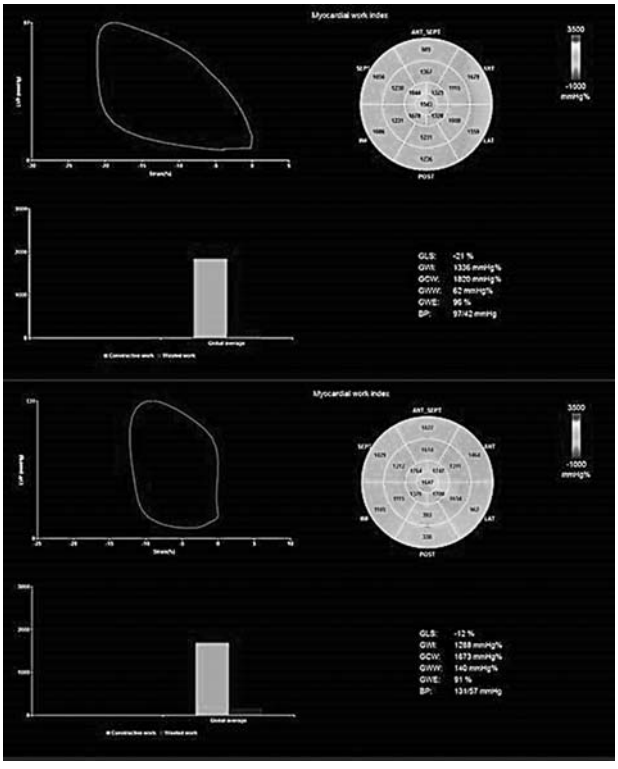
Introduction: Kawasaki disease (KD) is an acute vasculitis of unknown aetiology. It is associated with high morbidity and mortality due to the development of coronary artery aneurysms and myocardial dysfunction. The aim of this study is to evaluate sensitivity and specificity of left ventricular (LV) pressure–strain loop (PSL) area, which reflects regional myocardial work and metabolic demand, in predicting subtle myocardial abnormalities in KD patients with coronaries aneurisms.
Methods: A total of 47 patients (24 male, age 7.9 ± 8.09 years) were included in our study. Among the children admitted in our institution with a diagnosis of KD during the study time frame, 24 patients (16 male) with coronary artery dilatation (Z-score >2.5) were selected. These cases were compared with 23 (8 male) age-matched controls. Classical echocardiographic parameters of LV systolic function were normal for both groups, while global longitudinal strain (GLS) was decreased in 5 KD patients. Global work index (GWI) was calculated as the area of the LV PSL. From GWI, it was estimated also Global Constructive Work (GCW), Global Wasted Work (GWW) and Global Work Efficiency (GWE). We also made a subgroup analysis between KD patients with normal GLS (>-19) and control patients.



	KD group	CTR group	P	KD group (with normal GLS)	CTR group	P
GLS	-19.80 (-20.65 to -18.98)	-21 (-22.60 to -20.35)	0.006	-20 (-20.9 to -19.35)	-21 (-22.60 to -20.35)	0.07
GWI (mmHg%)	1332 (1179.75 - 1537.25)	1643 (1509-1763.5)	0.001	1336 (1207.5-1618.5)	1643 (1509-1763.5)	0.01
GCW (mmHg%)	1840 (1625.5-2094.25)	2017 (1848-2338)	0.04	1980 (1702-2163.5)	2017 (1848-2338)	0.25
GWW (mmHg%)	92 (57-136.25)	74 (59 – 127.50)	0.52	88 (55.5–131)	74 (59–127.5)	0.95
GWE (mmHg%)	93.50 (91.75–97)	96 (95 – 96)	0.15	95 (92.5–97)	96 (92–96)	0.60

Results: Despite normal LV systolic function by routine echocardiography, compared with controls, KD patients had a lower GWI (1332 [1179.75-1537.25] mmHg% in KDg vs 1643 [1509.00- 1763.50] mmHg% in CTRg, p=0.001) and GCW (1840 [1625.50- 2094.25] mmHg% in KDg vs 2017 [1848.00-2338.00] mmHg%, p= 0.04). However, there were not significant differences in GWW and GWE between the two group. When KD patients with normal GLS were analysed separately, they preserved a significant difference in GWI in comparison with controls (1336 [1207.5-1618.5] mmHg% in KDg vs 1643.00 [1509.00-1763.50] mmHg% in CTRg, p=0.01)
Conclusions: The estimation of myocardial work by PSL is a novel tool for the evaluation of patients with KD. GWI and GWE were significantly reduced in KD patients with dilated coronaries. Moreover, in KD patients with normal GLS, estimation of GWI may be a sensitive indicator of myocardial dysfunction and an adjuvant criterion to avoid delayed diagnosis of KD.

P-129

Variability and reproducibility of right ventricular longitudinal strain in patients with repaired Tetralogy of Fallot

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Introduction: (or Basis or Objectives): Right ventricular longitudinal strain (RVLS) has emerged as an approach for quantifying right ventricular function in diseases such as pulmonary hypertension and congenital heart disease. Data about the reproducibility of RVLS measurements, however, are missing. The aim of the current sudy were to analyze the reproducibility of RVLS on speckle-tracking echocardiography in the assessment of right ventricular function in patients with repaired Tetralogy of Fallot.
Methods: In this retrospective, single-center study, intra- and intra-observer intraclass correlation coefficients (ICC) were calculated. For this, 10 studies were randomly selected. These studies were analyzed by two independent observers and repeated by the same observer six months after the first analysis. Global longitudinal strain (GLS) and free wall strain (FWS) was evaluated in the four-chamber view using the standard semiautomatic method (Qlab software; Philips)
Results: An excellent inter-observer ICC was obtained, of 0.89 for GLS and 0.88 for FWS. In the same way, the intra-observer ICC was very good with an ICC of 0.96 for GLS and 0.95 for FWS. The mean ± SD inter-observer differences for the RV GLS was 2.5±1.8%.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusions: Our study shows that the intra- and inter-observer agreement for the RVLS are very good with differences < 3%. Therefore, longitudinal strain is a highly reproducible tool to assess right ventricular function in patients with repaired Tetralogy of Fallot.

P-130

Different CMR imaging modalities for native and patch-repaired right ventricular outflow tract: impact on percutaneous pulmonary valve replacement planning

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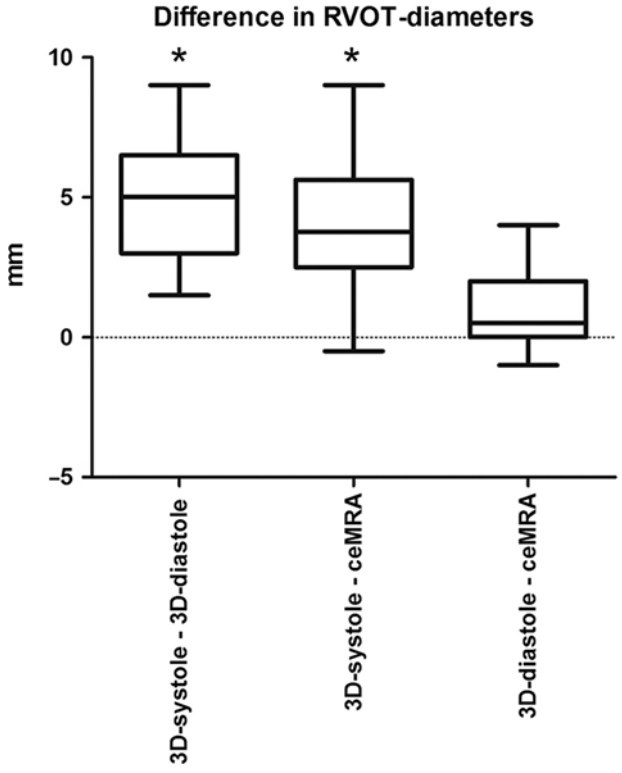
Introduction: Percutaneous pulmonary valve implantation (PPVI) in native or patched right ventricular outflow tract (RVOT), although still off-label, has proven to be feasible. The procedure is highly dependent on the size of the RVOT. The biggest percutaneous valve available in fact, measures 29 mm and in cases of excessive dilatation of the RVOT, PPVI may not be possible. Several methods exist to evaluate the size of the RVOT by cardiovascular magnetic resonance (CMR). We evaluated different CMR modalities for measuring RVOT diameters.
Methods: Thirty-one consecutive patients with native or patched RVOT were retrospectively evaluated. CMR was part of follow-up of patients with corrected Tetralogy of Fallot or pulmonary stenosis with significant pulmonary regurgitation (PR). CMR protocol included different sequences for the assessment of RVOT diameter, namely 3D-SSFP-whole-heart in systole, diastole and contrast-enhanced-MR-angiography (ceMRA). Diameters of the RVOT were assessed by the three sequences. The term RVOT is defined as the anatomic region from the right ventricular infundibulum to the branching of the pulmonary artery. Additionally, in patients who underwent cardiac catheterization (n=11) for PPVI, vessel diameters assessed by cine-angiography were compared to CMR.
Results: Systolic diameters of RVOT were significantly larger compared to diameters taken in diastole and ceMRA (median difference 5.0 mm and 3.8 mm). Diastolic and ceMRA diameters did not differ significantly. CMR diameters taken in systole showed no statistical difference to diameters taken by cine-angiography, while diastolic and ceMRA diameters were significantly smaller. PPVI was feasible to a maximal CMR diameter of 31 mm measured by SSFP-whole heart sequence in systole.
Conclusion: Absolute diameters of native RVOT differ depending on the CMR sequence and on timing of acquisition (systolic vs diastolic). Angiographic diameters best correlate to systolic CMR values. Data may help to select RVOTs suitable for PPVI.



P-131

Impaired pulmonary function and its association with clinical outcomes, exercise capacity and quality of life in children with congenital heart disease

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Background: Impaired pulmonary function is an independent predictor of mortality in adult congenital heart disease (CHD), but has been scarcely studied in the paediatric CHD population. This study aims to compare the pulmonary function of children with CHD to healthy controls, and evaluate its association with clinical outcomes, exercise capacity, and quality of life.
Methods: Cross-sectional multicentre study among 834 children (555 CHD and 279 control subjects) who underwent a complete spirometry and a cardiopulmonary exercise test (CPET). The 5th centile (Z-score = -1.64) was used to define the lower limit of normal. The association of clinical and CPET variables with spirometry was studied using a multivariate analysis. Children and their parents filled in the Kidscreen health-related quality of life questionnaire.
Results: Forced vital capacity (FVC) and forced expiratory volume in one second (FEV1) Z-scores values were lower in children with CHD than controls (-0.4 ± 1.5 vs. 0.4 ± 1.3, P<0.001 and -0.5 ± 1.4 vs. 0.4 ± 1.2, P<0.001, respectively), without any obstructive airway disorder. Restrictive pattern was more frequent in CHD patients than in controls (20% vs. 4%, P<0.0001). FVC Z-scores were predominantly impaired in complex CHD, such as heterotaxy (-1.1 ± 0.6), single ventricle (-1.0 ± 0.2), and complex anomalies of the ventricular outflow tracts (-0.9 ± 0.1). In multivariate analysis, FVC was affected by the age, the body mass index, the maximum oxygen uptake, the genetic anomalies, the number of cardiac surgery and cardiac catheter procedures. FVC and FEV1 correlated with self and proxy-related quality of life scores.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusion: These results suggest that pulmonary function should be monitored early in life, from childhood, in the CHD population.

P-132

Radial myocardial strain and biochemical markers of myocardial damage in children with hypertrophic cardiomyopathy

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Introduction: hypertrophic cardiomyopathy is a severely condition in children. Early detection of myocardial dysfunction allows to begin the treatment early in the disease.
Material and methods: 49 patients with hypertrophic cardiomyopathy (HCM) in age from 7 to 17 years (median 9 years) were examined by echocardiographic study. Radial myocardial strain was determined in 2D speckle tracking mode in 12 segments of the basal and medial part of the left ventricle (parasternal position, the short axis). Using the method of polynomial regression, we studied the interaction between: the radial deformation of the left ventricular myocardium, the thickness of the myocardial segment, troponin M and troponin I.
Results: Radial strain of the left ventricle myocardium ranged from 0.69 to 59.6%, the thickness of the myocardial segments varied from 6 to 43 mm. All children had asymmetric myocardial hypertrophy, more expressed in the anterior, anterior-septal basal and medial segments of the left ventricle. For myocardial segments with the thickness 4.24 Z-score or more, we discover decrease in radial deformation (less than 20%). In 15 children (30.6%) with number of hypertrophic segments from 3 to 7, was found an increase in biochemical markers of myocardial ischemia — troponin M ($M \pm 2\sigma = 0.19 \pm 0.1$ ng / ml) and troponin I ($M \pm 2\sigma = 0.76 \pm 0.1$ μg / l). The negative correlative relationship was obtained between these indicators and the radial deformation of the left ventricular myocardium ($r = -0.6$ and $r = -0.59$).
Conclusions: Changes in radial strain of the left ventricular myocardium in children with HCM reflect a systolic myocardial dysfunction, which is the basis for further observation and therapy.

P-133

Modern assessment of the arterial stiffness in adolescents with arterial hypertension

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Objectives: Condition of the vascular wall depends on many reasons, the main one is arterial hypertension (HTN). The aim of the study is to evaluate the parameters of the arterial stiffness in adolescents with HTN.
Methods: A group of adolescents from 12 to 17 years ($n = 340$) were divided into 2 main groups: 1 ($n = 248$) - children with stable hypertension (SH), following subgroups are allocated: with an increased body mass index (BMI), $n = 179$; with normal BMI, $n = 69$; with stage 1 HTN ($n = 124$) and stage 2 HTN ($n=124$). Control group ($n = 92$) included healthy children. In all patients we performed ABPM before the start of therapy. The following parameters were evaluated: aortic pulse wave velocity (PWVao), maximum rate of pressure rise (dP/dt max), ambulatory arterial stiffness index (AASI), arterial stiffness index (ASI).
Results: The mean values of PWVao, dP/dt max, AASI, ASI were significantly higher in the group with HTN (9.6 m/s, 1010.2 mm Hg/sec, 0.52, 154.05 mm Hg, respectively) compared to the control group (8.4 m/s, 678.3 mm Hg, 0.41, 138.5 mm Hg, respectively, p-value = 0.000).The dP/dt max and ASI indexes were more reactive than PWVAo, AASI and statistically higher in children with stage 2 HTN compared to patients with stage 1 HTN (156.05/152.05 mmHg, 1073.5/946.9 mm Hg/s, respectively, p-value = 0.03/0.000). All indexes have higher values in children with obesity, they also correlate with the grade of HTN (1/2) (PWVao: 9.8/9.9 m/s, AASI: 0.54/0.51, dP/dTmax (in HTN 2): 994.6/1092.5 mmHg/sec, p-value < 0.05 in all cases). In patients with stage 2 HTN ($n=124$) there are 25.5% adolescents ($n=31$) with LVMI >48 g/m2.7, they have a significant correlation with an increased PWVao (10.29 m/s, p-value = 0.000).
Conclusions: The use of non-invasive diagnostic method, ABPM, gives a highly informative assessment of parameters of arterial stiffness. Increased blood pressure is associated with increased vascular rigidity and target-organ damage (TOD). An important factor in the increase the rigidity of the vascular wall is the stage of HTN and level of BMI. Arterial stiffening may be considered as a prognostic factor for TOD.

P-134

Cardiac disorders in the treatment of infantile hemangiomas with propranolol

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Objective: Investigate the effect of propranolol on the cardiovascular system in the treatment of infantile hemangiomas.
Methods: A total of 132 children who started treatment with propranolol at the age of 1 month to 4 years with infantile hemangiomas of various localization were examined. The children underwent a complete cardiac examination, including a clinical examination, ECG, echocardiogram, Holter monitoring. The survey was conducted before the appointment of therapy with propranolol, every 3 months during therapy and after its cancellation.
Results: According to the echocardiogram, 6% of children initially diagnosed heart disease, (CHD, syndrome of noncompact myocardium of the left ventricle, PH). During the treatment, no child had a decrease in myocardial contractility. During treatment with auscultatory and according to ECG data, 1% of children had bradycardia. However, the Holter monitoring showed that all children after 3 months of treatment showed a decrease in heart rate during wakefulness by 9% of the norm (Me 130 ± 9.54). In 6% of children, during the treatment, rhythm pauses were identified, significantly exceeding the age norm (deviation rate 102-161%), which was an indication for changing the dose of the drug in the direction of its reduction. In one child, syncopal states were noted against the background of a pause in rhythm, which required discontinuation of therapy with propranolol. During treatment, AV block 1 degree was detected in 4% of children according to ECG data; according to Holter monitoring data, 18% of children showed AV block I – II degree. In 7% of children, AV blockade remained after discontinuation of propranolol therapy. Hypotension during treatment occurred in 11% of children.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusion: Clinical examination, ECG, echocardiogram, and Holter monitoring have made it possible to establish undesirable cardiovascular effects in 18% of children during therapy with propranolol. Holter monitoring before prescribing, during treatment and after termination allows to identify undesirable effects of therapy (bradycardia, cardiac rhythm pauses, AV-block II-III degrees) in time and adjust the dose of propranolol.

P-135

Von Willebrand Factor parameters, biomarkers for disease activity and Coronary artery lesion in Kawasaki disease

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Objectives: Von Willebrand factor (VWF), a large multimeric glycoprotein is essential for primary haemostasis. Increased plasma levels of VWF:Antigen (VWF:Ag) have been observed in cardiovascular diseases or vasculitis and are presumably caused by activation of the endothelium. The impact of Kawasaki disease (KD), a vascular inflammatory disease, regarding VWF:Ag, VWF:Collagen binding activity (VWF:CB) and VWF multimers structure analysis, has not been clarified yet. We investigated VWF-parameters in patients with Kawasaki disease (KD) as a surrogate marker for disease activity and Coronary artery lesion (CAL).
Methods: 28 KD patients, 10 with Coronary artery lesion were enrolled to this study. In 5 patients serial measurements were collected. VWF:Ag and VWF:CBA were determined by enzyme-linked immunoassay. The VWF:CB/VWF:Ag -ratio was calculated and the VWF structural features were assessed by multimer structure analysis. We evaluated the correlation between VWF parameters and standard inflammatory markers. The impact of the patients´ age, point of time of blood collection, being refractive to therapy and CAL on the VWF parameters was assessed. We furthermore analyzed the VWF-parameters predictive value for CAL.
Results: VWF:Ag and VWF:CB levels were significantly higher in the acute phase as compared to the convalescence phase. There was a moderate positive correlation (Pearson coefficient > 0,3) of all VWF parameters with CRP and VWF:Ag with a high platelet count. A lower VWF:CB/VWF:Ag-ratio was negatively correlated with a higher leucocyte count. Interestingly, the VWF:CB/VWF:Ag -ratio was significantly decreased in those patients with CAL (mean 0.96 vs. 0.64; p= 0.036) whereas the absolute levels of VWF:Ag and VWF:CBA did not show any differences with respect to CAE/CAA. Using a model to predict CAL, the AUC of the ROC was 0,84 (sensitivity of 60% and specificity of 94%). Those patients with very low VWF:CB/VWF:Ag-ratio in the acute phase had persistent CAL (1 year follow up).
Conclusion: Our study indicates that a comprehensive analysis of VWF-parameters may help to monitor KD inflammation and furthermore may help to detect those patients with increased risk for CAL. Further analyses should be performed in a larger study population.

P-136

Evaluation of Optical Coherence Tomography of the Pulmonary Arteries in Patients with and without Pulmonary Hypertension

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Introduction: To assess severity and prognosis of pulmonary hypertension (PHT), clinical evidence of right-ventricular failure, progression of symptoms, right ventricular function and hemodynamic parameters are taken into account, but predictions remain vague. We evaluated the imaging method of optical coherence tomography (OCT) of the pulmonary arteries for correlation with the severity of PHT in a group of pediatric patients.
Methods: OCT records ("runs") during right heart catheterization were performed with simultaneous angiography to confirm proper positioning of OCT catheter in the peripheral pulmonary arteries. 3 pictures of the best OCT run per patient were chosen to measure inner diameter of the vessel (VD) automatically and the wall-thickness (WT) at three different points manually. Mean VD and mean WT of each patient were correlated with mean pulmonary artery pressure (mPAP), and with pulmonary vascular resistance (PVR) and cardiac output (CO) both absolute and indexed. Wall appearance was studied in terms of layering and appearance of adventitial tissue.
Results: Of 20 Patients, 11 had mPAP 34-88, mean 55 mmHg, the controls had mPAP 8-18, mean 11 mmHg. 62 runs were performed with 1-8 in each patient. VD ranged from 0.88 to 4.87 mm (mean 2.80mm, median 2.58mm); WT ranged from 0.085 to 0.287 mm (mean 0.15mm, median 0.13mm). WT was increased in PH patients (0.18±0.06 mm vs 0.11±0.03mm, p=0.011) and correlated significantly with hemodynamics (mPAP, r=0.59; CO, r=-0.53; PVR, r=0.73 and PVRI, r=0.70, all p=0.009 or lower). There was a trend that appearance of the wall ('monolayer with an even network of vasa vasorum' versus 'multilayer disrupted by fibrous tissue') correlated with 'no or mild PH' versus 'severe PH'.
Conclusion: Wall thickness and wall appearance of the distal small pulmonary arteries as imaged by OCT in patients with PHT correlates with PHT as such and reflects severity of PHT. Further studies appear to be justified to evaluate if information from OCT can contribute to medical care of children with pulmonary hypertension.

P-137

Comparing wall shear stress values by using different 4D flow postprocessing tools in cardiovascular magnetic resonance in bicuspid aortic valve disease

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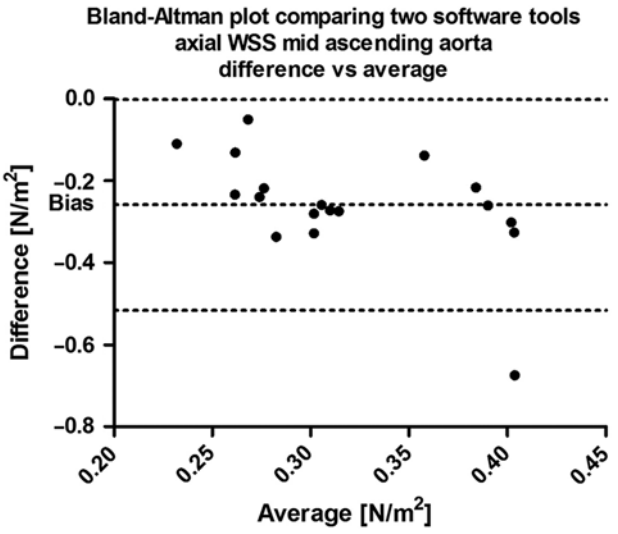
Introduction: Over the last ten years 4D-flow in cardiovascular magnetic resonance has been established as a valid method to visualize hemodynamic flow patterns in congenital heart disease. During that period various tools for flow visualization and wall shear stress (WSS) evaluation have been improved constantly. In terms of reproducibility it is important that different software will calculate comparable results. The aim of this study is to evaluate differences

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



in WSS estimation by comparing results generated in 2008 and recalculated 2018 based on the same data by reassessment with newer scientific 4D software.

Methods: Eighteen complete 4D flow datasets of patients with bicuspid aortic valve disease were evaluated for WSS in 2008 and reevaluated in 2018 by using MEVISFlow® (MEVISFlow®; Fraunhofer Institute for Medical Image Computing, v10.3, Bremen, Germany). In 2008 all values were generated with EnSight® and dedicated software based on Matlab® (EnSight®; CEI, Apex, NC, USA; The MathWorks, Natick, MA, USA). All data were analyzed by two skilled persons.

Results: Data analysis showed slight, but significant differences in WSS measurements depending on the software used for the evaluation. The mean difference (old vs. new) of the axial WSS at the level of the mid ascending aorta was -0.26 N/m2± 0.13 SD (p= 0.0002), the circumferential WSS differed by mean -0.25 N/m2 ± 0.17 SD (p=0.0003) and the magnitudinal WSS showed a difference of mean -0.14 N/m2 ± 0.19 SD (p=0.0035). Bland Altman analysis showed a 95% limit of agreement of -0.52 to -0.003 (Bias -0.26; Figure), -0.58 to 0.09 (Bias -0.25) and -0.51 to 0.23 (bias -0.14), respectively. Evaluation by the two different observers showed no difference (mean difference 0.02 N/m2; 95% confidence interval -0.002 to 0.041).

The new postprocessing software systematically generated slightly higher WSS parameters.

Conclusions: Different software tools for WSS assessment by 4D flow may generate different values. In our study we got slightly higher values by using a newer tool. For longitudinal 4D flow data evaluation it will be important to use the same postprocessing tools.

P-138

Evaluation of different coronary artery Z-Score models in a German population based study cohort of 310 patients

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Objectives: Kawasaki disease (KD) can be complicated by Coronary Artery Aneurysms (CAA). Diagnostic accuracy of CAA significantly influences subsequent diagnostic and therapeutic decisions. Today, 7 different Z-Score models, standardising the numerical measurement of the Coronary artery diameter's, have been established from measurements of healthy North American and Asian children. These scores have however never been uniformly evaluated in a representative KD patient cohort. In addition, no recommendation exists which score may be best useful in Western Europe.

Methods: We recruited from the active population-based German Pediatric Surveillance Study (ESPED) for KD and from the Ludwig-Maximilian-University of Munich echocardiographic patient cohort. Echocardiographic reports performed during the acute phase of KD were systematically screened regarding specifications of coronary artery diameters. All patients evaluated fulfilled the AHA diagnostic criteria for KD. From all available coronary artery diameters, Z-Scores were calculated according to all available models. The Z-scores were compared for normal distribution and stability concerning the diagnosis of coronary artery ectasia (CAE, Z-Score >2 and <= 2,5) and coronary artery aneurysm (CAA, Z-Score > 2,5).

Results: In 310 out of 408 echocardiografic reports at least one coronary artery diameter was reported. In all but 5 reports of those without given coronary artery dimensions, coronary arteries were judged as normal by the treating physician. Applying the different Z-score models to the documented measurements, the percentage of CAA varied considerably; LMCA 25 – 45%, RCA and LAD 30 – 39% respectively. CAE was less frequently diagnosed and therefore showed less variance (LMCA 8-11%, RCA 2-6%, LAD 1,5 – 6%). The Kobayashi- and Dallaire Z-Score models showed the most concordant results among all Coronary arteries, these scores however revealed higher rates of CAA than the others.

Conclusions: There are considerable discrepancies between the different Z-Score models available in terms of determining CAA and CAE. Since the Z-Score models proposed by Kobayashi and Dallaire, obtained comparable results to the investigators here and higher rates of CAA, we would propose to use either one of these models. Underdiagnosis and delayed treatment initiation or exacerbation might be prevented that way.

P-139

Utility of Pulse Oximetry Screening of Newborns for Non-critical Cyanotic Congenital Heart Diseases

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Background: Several large randomized trials have shown that predischarge screening of newborns by pulse oximetry helps in early diagnosis of cyanotic congenital heart diseases (CCHD). Most studies have used a cut off value of oxygen saturation (SaO2) of 95% or more to exclude CCHD. However newborns with non patent ductus arteriosus (PDA) dependent CCHD (non-critical CCHD) may have a SaO2 of >95%, thereby passing the oximetry test. This may falsely reassure the parents.

Objective: We aimed to study the SaO2 difference between the critical (PDA dependent) and non-critical CCHD in newborns.

Methods: It was a cross sectional observational study conducted over a period of three years in a community hospital. All babies born during a specific 8 hour period of the day were recruited over a period of 3 years. The investigations included routine clinical examination, post ductal pulse oximetry (within 48 hours of life) and echocardiography (to screen for congenital heart disease) for all babies.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Results: Among the 20307 newborns screened, 1298 were excluded due to non availability of total data. Echocardiography detected CCHD in 33 newborns, giving a prevalence of 1.7/1000 (95% CI: 1.2-2.4/1000). Of these, 23 had critical CCHD and 10 had non critical CCHD (tetralogy of Fallot or its variant: 5; non-obstructive total anomalous pulmonary venous drainage: 2; common arterial trunk: 2; double inlet left ventricle with pulmonary hypertension: 1). Overall, SaO2 was higher in non-critical CCHD as compared to critical CCHD (94%±2.05% vs. 82.9% ±14.3%, p=0.001). A SaO2 of <95% was present in 21/23 (91%) newborns with critical CCHD and in 6/10 (60%) newborns with non-critical CCHD. If a cut off value of < 92% was considered, 9/10 (90%) of non-critical CCHD were missed by pulse oximetry as against 9/23 (39%) critical CCHD.
Conclusion: Pulse oximetry is a sensitive tool for detecting CCHD, however 40% of non-critical CCHD may be missed if SaO2 cut off of <95% is used. The non-critical CCHD could be complex with a suboptimal long term outcome, requiring parental counselling.

P-140
The role of regular physical activity in Fontan Circulation
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Introduction: Functional capacity in Fontan circulation is commonly impaired. Cardiopulmonary exercise test (CPET) variables are prognostic in these patients. We sought to undertake a regular CPET evaluation of Fontan patients followed in outpatient clinic at a tertiary hospital and find predictors of functional capacity.
Methods: Forty-four Fontan patients followed in outpatient clinic were prospectively evaluated with CPET and transthoracic echocardiogram (TTE). Regular physical activity was defined as extracurricular organized sports participation or at least two 45-minute periods of physical activity each week. Regarding atrioventricular (AV) dominant valve regurgitation on TTE, patients were classified by two experienced operators in 2 groups: those without significant regurgitation (none or mild) and those with significant regurgitation (moderate or severe). Statistical inference was performed using R CRAN version 3.5.0. Linear regression and ANOVA were used for continuous variable correlation, Chi-squared test and logistic regression were used for binomial variable correlation.
Results: Median age at Fontan completion was 6 years (SD 3) and median age at current evaluation was 19 years (SD 7). Sixteen patients (36%) had regular physical activity. Mean peak oxygen uptake (peak VO2) was 27.6 ml/kg/min (SD 6), peak VO2 as percentage of predicted value was 67% (SD 15), VO2 at ventilatory threshold was 16 ml/kg/min (SD 3), VE/VCO2 slope was 36.7 (SD 7), respiratory exchange ratio (RER) was 1.06 (SD 0.08), maximal heart rate (HR_max) was 164 bpm (SD 26) and peak oxygen saturation was 87% (SD 8). Peak VO2 was inversely correlated with age (p=0.006) and significant AV regurgitation (p=0.003) and had a strong positive relation with the practice of regular physical activity (p=0.001). Multivariate analysis showed that higher peak VO2 was independently associated with regular physical activity and absence of significant AV regurgitation ($R2 = 0.327$, F test with $p = 0.001$).
Conclusion: Peak VO2 is a marker of functional capacity and prognosis in Fontan patients, usually impaired. Lower peak VO2 was related with significant AV regurgitation. We identified a strong positive relation between peak VO2 and regular physical activity, which is relevant for patient guidance and recommendations.

P-141
Atrioventricular regurgitation and biomarkers in Fontan Circulation
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Introduction: The detrimental role of the Fontan circulation in the hepato-intestinal system has been well documented. Fontan patients are at risk of complications such as hepatic fibrosis and protein losing enteropathy (PLE). Our goal was to understand the relationship of possible disease biomarkers to other aspects of this circulation, detected by echocardiography.
Methods: Forty-four Fontan patients followed in outpatient clinic were prospectively studied with laboratory evaluation and transthoracic echocardiogram (TTE). Regarding atrioventricular (AV) dominant valve regurgitation on TTE, patients were classified by two experienced operators in 2 groups: those without significant regurgitation (none or mild) and those with significant regurgitation (moderate or severe). Selected biomarkers were related to hepatic disease (aspartate transaminase (AST), alanine aminotransferase (ALT), total bilirubin (TB), gamma-glutamyl transferase (GGT) and alkaline phosphatase (ALP)), PLE (total protein and albumin), intestinal inflammation (fecal calprotectin (FC)) and cardiac function (NT-proBNP). Statistical inference was performed using R CRAN version 3.5.0. Linear regression and ANOVA were used for continuous variable correlation.
Results: Median age at Fontan completion was 6 years (SD 3) and median age at current evaluation was 19 years (SD 7). No patient had previous diagnosis of hepatic disease and 4 patients (9%) had PLE. Mean values and standard deviation (SD) of selected biomarkers were: total protein 7g/dl (SD 1), albumin 4.5g/dl (SD 0.7), AST 30U/L (SD 10), ALT 28U/L (SD 11), TB 0.9mg/dl (SD 0.5), GGT 78U/L (SD 62), ALP 148U/L (SD 101), FC 92mg/kg (SD 62), NT-proBNP 388pg/ml (SD 1031). The most common abnormality on TTE was significant AV regurgitation in 20 patients (45.5%). Statistical analysis showed a strong correlation between this and levels of TB (p=0.016), GGT (p=0.013), FC (p=0.003) and NT-proBNP (p=0.045). Higher FC was also related to higher NT-proBNP (p=0.004).
Conclusion: Fontan patients with significant AV valve regurgitation have abnormal values of several biomarkers related to cardiac, hepatic and intestinal disease. Presence of more than mild AV valve regurgitation in Fontan patients defines a high-risk group requiring tight follow-up.

P-142
Role of the global longitudinal strain on early diagnosis of anthracycline induced cardiac dysfunction in paediatric patients with acute lymphatic leukaemia
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Introduction: Paediatric patients with acute lymphoblastic leukaemia (ALL) are at high risk of developing cancer therapeutics–related cardiac dysfunction (CTRCD). Global Longitudinal Strain (GLS) has been used as an early predictor of late cardiac dysfunction. The objectives of this study are to demonstrate whether the GLS is feasible in the paediatric age, if it changes before the ejection fraction

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

(EF) and to define whether a therapeutic action based on GLS is useful.
Methods: Seventy-seven patients diagnosed with ALL and treated with the Italian AIEOP-BFM ALL 2009 protocol were enrolled. All patients did an echocardiography evaluation at baseline and before every cycle of anthracycline. In our department there are two different follow up clinics: the first follows the new ESC guidelines and uses the GLS in addiction to LVEF to estimate LV function (group 1), while the second follows the AIEOP protocol where the EF is the only parameter required (group 2). A value of LVEF <10% from baseline to the lowest normal limit and a GLS> -19% were considered pathologic. GLS of patients in group 2 was calculated from the apical 4 chamber view retrospectively. For every patient three exams were considered: the baseline, the nadir of GLS and stop therapy. Patients with demonstrated cardiac toxicity were treated with carvedilol.
Results: Forty-four patients (57%) terminated the two years cancer therapy and 41 completed all the echocardiography assessments. 41 pt were in the first group and 36 in the second group. Overall, none developed CTRCD as per EF, while 19 pt developed a GLS> -19% during the follow up. The EF decreases in both groups between baseline and nadir and remained stable between nadir and stop therapy. The GLS had a similar path in the first phase, but it improved between nadir and stop therapy only in the first group, in which carvedilol was started.
Conclusions: CTRCD is relatively frequent during treatment for ALL in childhood. GLS seems to be feasible in the paediatric age and an early marker of subclinical cardiac dysfunction. In the case a beta-blocker is started in response to a pathologic GLS, an improvement was noticed.

P-143
Ventricular septal defect associated with aortic valve prolapse: an echocardiographic study

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Objectives: Ventricular septal defects (VSD) can be associated with aortic valve prolapse (AVP) with or without regurgitation. Aortic valve prolapse may be due to both lack of leaflet support and Venturi's phenomenon. There is a great diversity in the literature about the anatomic type of the VSDs associated with AVP. The objective of the study was then to assess the anatomy of the VSD on echocardiography.
Methods: From 2009 to 2017, 51 consecutive patients with VSD complicated by AVP were admitted for surgery. We screened retrospectively all preoperative echocardiographies in double-blind test. We analyzed the anatomic type of the VSD according to ICD-11 classification and looked for outlet septum malalignment, the nature of the prolapsed aortic leaflet, the severity of aortic regurgitation and other complications (subaortic membrane, right ventricular stenosis).
Results: The VSD was central perimembranous (pm) in 23 patients (45%), outlet with malalignment in 21 (41%) and outlet juxta-arterial in 7 (13.7%). We found a significant difference between the initial diagnosis and the final one after reviewing echography in double-blind test: among the 23 VSD initially described as central pm, eleven were reclassified as outlet VSD, and one outlet VSD was reclassified as central pm ($p<0.001$). Aortic valve prolapse involved the right aortic cusp in 98% of patients. Aortic regurgitation was severe in 2 patients, moderate in 5, mild in 24 and trivial or absent in 20. Outlet VSDs were significantly more often associated with aortic regurgitation ($p=0.036$). Subaortic membrane was found in 8 patients (15.6%), 6 with outlet and 2 with central pm VSD (p:0.001). Right ventricular obstruction was found in 8 patients (15.6%).
Conclusion: Aortic valve prolapse complicates indifferently central pm and outlet VSD. However, aortic insufficiency is more frequent in outlet VSD. The accurate echocardiographic diagnosis of the anatomic type of the VSD is difficult and multiple views are necessary.

P-144
4D flow MRI to assess right ventricular outflow tract in patients undergoing transcatheter pulmonary valve replacement for severe pulmonary regurgitation

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Magnetic resonance imaging (MRI) is essential to assess right ventricular enlargement and function as well as the severity of pulmonary regurgitation before pulmonary valve replacement. Transcatheter pulmonary valve replacement (TPVR) has emerged as an alternative to surgical replacement but limited by the size of the right ventricular outflow tract (RVOT) especially in case of isolated PR.
The aim of the study was to compare the measurements of native RVOT between different MRI imaging sequences and catheterization findings
Methods: Single-center retrospective study of patients who underwent TPVI for severe pulmonary regurgitation assessed by 4D flow MRI, without significant residual RVOT obstruction between March 2015 and January 2017. Balloon calibration was used as the reference.
Results: Sixteen adults with repaired tetralogy of Fallot were included (mean age: 39.9 +/− 12.9). 1 patient had a too large RVOT to achieve a TPVR, 1 patient had TPVR with Edwards XT 29mm valve, 2 patients had classical Melody 22m valve implantation procedure and the others had various techniques for Melody valve implantation in large RVOT (jailing or unconventional method). Mean RVOT measured 24.7+/− 3.2mm by balloon calibration, 24.2 +/− 3.3 mm and 22.3 +/− 3.0mm by 4D flow MRI respectively in systole and diastole, and 21,4+/− 2.9mm in 2D SSFP MRI. No statistical differences were found comparing differents measurements dimensions. Good correlation was found between balloon calibration and MRI 4D flow in systole ($r= 0.78$, $p=0.006$), while no correlation existed between balloon calibration, 2D MRI and 4D flow MRI in diastole. Balloon calibration was also well correlated to angiograms measurements ($r = 0.89$; $p < 0.0001$ and $r = 0.79$; $p < 0.0001$ respectively).
Conclusion: 4D flow MRI sequence allowing 3D measurement through the cardiac cycle, appears as an interesting technique to measure RVOT in daily practice and therefore to plan TPVI.

P-145
Clinical manifestation and outcomes of children with hypertrophic cardiomyopathy in Kosovo

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

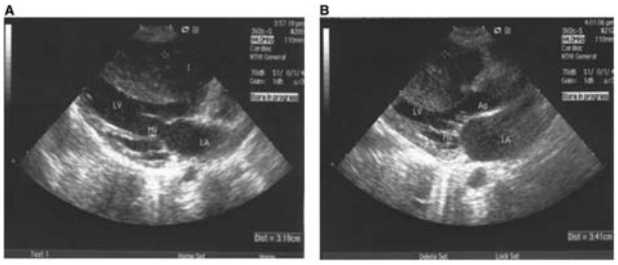

Figure 1.
Parasternal left ventricular long-axis echocardiografic section obtained a patient in diastole [A] and systole [B] with hypertrophic cardiomyopathy. LV- left ventricle, LA- left atrium, Ao- aorta, MV- mitral valve.

Introduction: Identification of the manifestations, assessment and fellow up of children with hypertrophic cardiomyopathy (HCM) by transthoracic echocardiography may be important for clinical management and our understanding of the pathogenesis.
Aim: of presentation is to present all children diagnosed with HCM in Kosovo, during the period 2005 -2017, diagnosed by transthoracic echocardiography, treatment and outcomes.
Result: Here we present a comprehensive analysis of 43 patients seen in Kosovo, of whom 23 were male, aged between 4 months and 9 years at first presentation, (median of 2 years and 3 months). Cardiac failure, seen in almost half of them, was the most frequent presenting feature. In admission, on the chest x-ray, the cardiothoracic ratio was increased, to a mean of 72% in 5 infants and to 65% in 37 older children. Measured by transthoracic echocardiography, in 28 patients hypertrophy of left ventricle was asymmetric while 15 had concentric hypertrophy. Left ventricular ejection fraction was depressed in the 21 patients. Patients in cardiac failure received various combinations of diuretics, B-blockers, ACE inhibitors and aspirin. Death occurred in 8 patients, in 4 of them shortly after admission, 3 left Kosovo and continued examination abroad, and the remaining 32 were followed- up for a mean 42 months, with a range from 5 to 115 months. Surgical intervention was not performed to none of them, despite of clinical and echocardiographic indications. Recovery was noted in 14 patients but still requiring anti-failure medications. Slightly over two-fifths died. Of those with asymmetric form, 45% died, in half of those presenting in infancy, and 89% of those who presented at admission with signs of cardiac failure.

Key words: hypertrophic cardiomyopathy, left ventricular hypertrophy, heart failure, myectomy transthoracic echocardiography.

P-146
Poor agreement between echocardiographic and CMR derived peak strain parameters in patients with repaired Tetralogy of Fallot

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Table 1: Agreement between Echocardiographic and CMR Peak Strain Parameters

Peak Strain Parameters	*Mean bias (%)*	*Limits of agreement (%)*	*CoV (%)*	*Intraclass correlation Coefficient*
LV- Global Longitudinal Strain (n = 37)	0.23	-9.15 - 9.60	25.7	0.271
LV- Global Circumferential Strain (n = 36)	1,17	-11.91 - 9.56	23.9	0.377
RV- Global Longitudinal Strain (n = 35)	0.58	-12.00 - 13.16	32.0	0.110

Abbreviations: CoV: Coefficient of variation, CMR: Cardiovascular magnetic resonance, LV: left ventricular, RV: right ventricular.

Objectives: Patients with Tetralogy of Fallot (TOF) are at risk for deterioration of ventricular function. Echocardiography and cardiovascular magnetic resonance imaging (CMR) are used to assess ventricular function. Both imaging modalities can assess myocardial deformation (strain), which has been linked to cardiac outcome. This study aims to: 1) describe echocardiographic-strain and CMR-strain parameters and their reproducibility using the same post-processing software, 2) investigate the agreement between echocardiographic-strain and CMR-strain and 3) investigate the potential correlation between strain and other CMR parameters.
Methods: In a prospective multicentre study patients underwent a CMR and echocardiography on the same day. Echocardiographic-strain and CMR-strain of the left ventricle, LV (longitudinal and circumferential) and right ventricle, RV (longitudinal) were analysed with the same post-processing software (TomTec), which contained a new dedicated RV algorithm. Inter-observer agreement and agreement between echo-strain and CMR-strain was evaluated using Bland-Altman analysis, coefficient of variation (CoV) and interclass correlation coefficient. Correlations between strain and CMR parameters were evaluated using the Spearman's rank or Pearson's correlation coefficient.
Results: We included 40 TOF patients, 27 (72%) male. The median age was 18.8 years (IQR: 15.4-24.2), time after TOF repair 18.1 years (14.8-23.2). The agreement between global peak echo-strain and CMR-strain parameters is shown in Table 1. A significant difference in peak RV global longitudinal (RVGLS) derived from echocardiography and CMR was found (-21.3±4.9 vs -24.4±4.8, p=0.007). The inter-observer CoV for RV and LV peak echo-strain and CMR-strain ranged between 9.8-17.9% and 9.3-15.2% respectively. CMR derived LV and RV ejection fraction (EF) correlated significantly with echocardiographic LV global circumferential strain (r=-0.36, p=0.03 and r=-0.51, p=0.001) and LV global longitudinal strain (r=-0.41, p=0.01 and r=-0.47, p=0.003). Echocardiographic RVGLS was significantly associated with LVEF (r=-0.36, p=0.03) but not with RVEF. CMR RVGLS was significantly associated with RVEF (r=-0.46, p=0.01) and LVEF (r=-0.36, p=0.03)
Conclusions: We report that the agreement between echo-strain and CMR-strain parameters in a cohort of TOF patients is weak, despite using the same post-processing software with a dedicated RV measurement tool. This limits the ability to interchange these imaging modalities in the follow-up of individual patients.

P-147
Renal dysfunction and aortic stiffening in the children with congenital heart diseases

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Background: Arterial stiffening may decrease impedance mismatch in the proximal arterial system which transmits blood pressure directly to the micro vessels, leading to microvascular damage

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

and renal dysfunction. Renal dysfunction may, in turn, attribute to vascular dysfunction through hypertension, abnormal glucose/lipid metabolism, inflammation, oxidative stress and activation of the renin–angiotensin system. That renal dysfunction correlates with increased arterial stiffness has already been described in adults. We hypothesized that a similar reno-vascular interaction also exists in children with congenital heart disease (CHD).
Methods and results: During cardiac catheterization, pulse wave velocity (PWV), as a marker of arterial stiffness, was measured during the catheter drawback from ascending aorta to femoral artery in 200 children (age, 3.4±4.5 years) with various CHDs, including functional single ventricle (n=117) and repaired/unrepaired biventricular heart (n=83). Relationship between PWV and estimated glomerular filtration rate (eGFR: 112±27, 38-180 ml/min/1.73m2) derived from serum level of creatinine (0.31±0.12, 0.16–0.88 mg/dl) was then investigated. The PWV (mean±SD) of the proximal and distal aorta were 601±222 and 442±142 cm/sec, respectively. The PWV-proximal aorta but not PWV-distal aorta correlated significantly with eGFR (PWV= 920–2.48*eGFR, $p<0.05$). Multivariate linear regression analysis confirmed eGFR as an independent determinant of PWV-proximal aorta ($p=0.028$) even after controlling for confounding factors (age, sex, blood pressure, and disease type), whereas age was markedly associated with PWV-distal aorta ($p=0.031$). Interestingly, PWV-distal aorta was positively correlated with serum levels of procollagen type-III peptide even after adjusting for age. Among disease groups, tetralogy of Fallot was also an independent determinant of high PWV ($p=0.0011$).
Conclusions: This is the first report of reno-vascular interaction even in children with CHD and relatively preserved renal function. Because increased arterial stiffness is an independent risk factor for cardiovascular disease, and because chronic kidney disease is an independent risk factor for cardiovascular morbidity and mortality, these results highlight the importance of close follow-up of reno-vascular function and its interaction with a special consideration of disease specificity in children with CHD. Studies on the effects of reno-vascular protective treatments on prognosis are warranted.

P-148
Cardiopulmonary Exercise Test (CPET) usefulness for decision making in patients with repaired Tetralogy of Fallot (ToF) and secondary severe pulmonary regurgitation (PR)

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Background: Patients with severe PR have significantly dilated right ventricle (RV). Its progressive dysfunction and increased volume overload may impact in exercise capacity. The aim of this study is to compare changes in CPET in these patients and consider the implications of these changes.
Methods: Retrospective case-control study was lead in our Paediatric Cardiology Division at Donostia Hospital, comparing severe PR patients secondary to ToF and healthy subjects. All underwent for standardised CPET, using Godfrey ramp protocol as recommended by the European Society of Cardiology (ESC). Measured variables were: work rate (WR), predicted peak oxygen consumption (peak VO2), oxygen uptake efficiency slope (OUES), ratio of minute ventilation to carbon dioxide production (VE/VO2 slope), peak heart rate (pHR) and rate-pressure product (RPP) were registered. Mann Whitney test was used to compare continuous variables and Chi-square test for categorical changes.
Results: Twenty patients (11 girls, 55%) with previous repaired ToF and secondary severe PR (group A, mean age 13,5 ± 2,8 years) were compared to twenty sex-matched healthy teens (group B, mean age 12,2 ± 2,2 years; p=0,1). Group A raised a significantly lower WR (110,3 ± 8,6 vs 125,8 ± 6,3 watts, p=0,04). In the same way, group A had a significantly lower predicted peak VO2 (81,1 ± 5,1 vs 97,8 ± 2,9%; p=0,017) as well as a lower OUES (1,8 ± 0,1 vs 2,5 ± 0,1; p=0,045), a lower pHR (165 ± 4,7 vs 182,6 ± 2,1 beats per minute; p=0,02), and a lower RPP (20589 ± 1053 vs 34017 ± 7251 beats per minute per mmHg; p= compared to group B. On the other hand, VE/CO2 slope was significantly higher in group A (31,6 ± 1,3 vs 27,6 ± 1; p= 0,04).
Conclusion: The results of CPET showed that patients with repaired ToF and secondary severe PR had a low maximal exercise capacity, even among asymptomatic patients, suggesting impaired exercise capability. CPET should be considered to assess these patients functional capacity in order to assess the right pulmonary valve exchange timing.

P-149
Echocardiographic follow up of perinatally HIV-infected children and adolescents: results from a single center retrospective cohort study in Brazil

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Introduction: The effects of HIV and antiretroviral therapy on the cardiovascular system of perinatally infected children and adolescents are not fully understood. Most echocardiographic studies are based on a single evaluation of each patient, which precludes the precise analysis of what happens throughout their development. Objective: to determine the prevalence of cardiac abnormalities in a retrospective cohort of perinatally HIV-infected patients and to investigate associations between echocardiographic and clinical data during their long-term follow up.
Methods: Medical records from 148 perinatally HIV-infected patients (0-18y) seen between 1991 and 2015 were reviewed. Echocardiographic data included the presence of right and/or left ventricle dilation (diastolic diameter z-score > +2), septum and/or LV wall hypertrophy (z-score > +2), LV systolic dysfunction (EF < 55%) and pulmonary hypertension (PASP >35 mmHg).
Results: 480 echocardiograms were analyzed and 46 (31.1%) patients showed cardiac abnormalities. Only 6 (1.2%) echocardiograms were accompanied by heart failure symptoms. Nadir CD4 count was lower in patients with abnormal echocardiogram: 202 (5-1746) vs. 263 (4-1485) cells/μl, $p = 0.02$. RV dilation was present in 28/148 (18.9%) patients, transient in 15/28 (53.5%) and associated to CDC category C (66% vs. 44,2%; $p = 0.002$). LV dilation was present in 32/148 (21.6%) and was transient in 14/32 (43.7%). It was associated to lower absolute CD4 count [536 (10-1390) vs. 590 (5-5962) cells/μl; $p = 0.048$] and lower duration of HAART [1.5 (0-16) vs. 4.7 (0-16) years; $p = 0.0001$]. Pulmonary hypertension was detected in 13/148 (8.7%) patients, transient in 6/13 (46%), associated to viral load log > 5 (26.3% vs. 8.3%; $p = 0.021$) and to opportunistic infections (31.6% vs. 4.5%; $p = 0.0001$). The absence of protease inhibitors in therapeutic regimen was associated to LV dilation (69.5% vs. 43.3%; $p = 0.0001$), RV dilation (63.3% vs. 45.6%; $p = 0.01$), LV wall hypertrophy (83.3% vs.46.9%; $p = 0.013$) and LV systolic disfunction (88% vs. 45.6%; $p = 0.0001$).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusions: Echocardiogram detected subclinical cardiac abnormalities, that were transient in almost half of patients. Data suggests that immunologic status and therapeutic strategies can influence cardiac disease burden of perinatally HIV-infected children and adolescents.

P-150
Analysis of association of ventricular strain parameters with LV mass index in children with remodeling LV after heart transplantation

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Objectives: The differences in body surface area (BSA) between recipients and donors of heart transplantation (HT) are common in children. In this study, we investigated the association of the LV (left ventricle) mass index with the myocardial longitudinal strain (LS) over time up to 1 year after heart transplantation in children.
Methods: We reviewed 81 consecutive patients under 18 years old who received the heart between August 1997 and June 2017. 29 patents had the donor BSA of more than 130% of the recipient's BSA. 16 of them had echocardiographic record in which we could analyze LV mass index and the strain. Also, we analyzed 19 patients with echocardiographic results whose BSA was less than 130% of donor's BSA. 3 analyzes were performed over a one-year period. The first, second and third analysis was done at 1.1±0.5, 4.9±1.7, 12.4±2.7 months after HT, respectively. Echocardiographic results obtained within 1 month after a diagnosis of acute rejection were excluded
Results: The study included 35 patients underwent HT. In the group of patient's BSA was greater than 130% of donor's, the mean LV mass index for each period was 147.1±53.7, 132.2±37.0, 110.4 ±34.4 g/m2, which tend to decrease with time after HT as ventricular remodeling occurs (p=0.061). In the other group, the mean LV mass index for each period remained same (p=0.614). The LS of LV negatively increase -12.2±3.3, -14.4±5.1, -17.4 ±5.0 over time in 4 chamber view in the group of BSA ratio ≥ 1.3 (p=0.011) whereas there was no change in the other group (p=0.196). Although the LV mass index and the longitudinal strain of LV did not correlate with each other until the second analysis, they were significantly correlated with each other in the third analysis (p=0.042) in the group of patients with big BSA ratio.
Conclusions: The increasing tendency of LV LS of 4 chamber view was shown in children with remodeling process of LV only in children with BSA donor/recipient ratio ≥ 1.3. The association of LS and mass index of LV was clear only in patients with big BSA ratio after the 1 year from HT.

P-151
Cardiac Mechanics Comparison between pmVSD post Percutaneous Transcatheter Closure

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Objective: Percutaneous transcatheter closure of perimembranous ventricular septal defect (pmVSD) with occluder has been most widely used in China. In this study, we aimed to analyze ventricle performance post percutaneous transcatheter closure of pmVSD.
Methods: 40 (21 males) pmVSD pediatric patients (age 6.0 ± 1.2 years) and 40 (25 males) healthy controls (age 5.8 ± 1.1 years) were recruited. All subjects were studied with conventional and tissue Doppler echocardiography. Strain and strain rate of left ventricle (LV) and right ventricle (RV) were assessed by 2D speckle tracking echocardiography (2D-STE).
Results: Mean diameter of pmVSD was 3.82 ± 0.59 mm, mean diameter of pmVSD occluder was 6.3 ± 1.0 mm, and mean time post percutaneous pmVSD closure was 3.22 ± 0.78 years. No significant differences were observed in LV eject fraction, RV Tei index and transatrioventricular velocity (E and A) 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 provide valuable information on the quantitative assessment of ventricular function in children post percutaneous transcatheter closure of pmVSD. And it appears that percutaneous closure of pmVSD is safe and effective in selected pediatric patients.

Keywords: perimembranous ventricular septal defect, 2D speckle tracking echocardiography, strain, strain rate.

P-153
Implementation of 3D segmentation imaging and 3D printing in the clinical practice

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Background: 3D segmentation imaging and 3D printing have become a valuable tool for the preoperative preparation and decision making. In particular with complex CHDs in which it is difficult to represent specific and complicated 3-dimentional intra-cardiac and vascular relations via conventional echocardiography or CT/MRI. The method also provides aid in the communication with the patient/parents and the education of personnel and students.
Objective: To examine and verify the benefits of the method in the preoperative preparation. To explore the patients/diagnoses which would be indicated for this form of advanced imaging. To implement the method in the clinical practice of our center.
Method: 3D segmentation was performed in 9 cases during 2017 and 2018. 5 cases of Double aortic arch(DAA), 1 case of Pulmonary atresia, VSD and MAPCAs, 1 case of Common arterial trunk, 1 case of Tetralogy of Fallot, 1 case of Superior sinus venosus ASD with PAPVR. Segmentation was achieved via "3D slicer" software on data from contrast CT performed on the patients. 3D printing was performed in 4 of the above mentioned cases – 3 cases of DAA, 1 case of pulmonary atresia, VSD and MAPCAs. The models were printed on a generic FDM 3D printer, and in 1 case of DAA the model was printed on "Ultimaker 3" in PLA plastic. All models were compared to the intraoperative findings, the original imaging study and for the case of pulmonary atresia, VSD and MAPCAs – with the post-mortem anatomic specimen.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Results: There is a clear overlap between primary imaging, segmented image, printed model and the intra-operative/post-mortem findings and dimensions.
Conclusion: 3D segmentation imaging and printing is a useful method which provides in-depth imaging of complex anatomical relations. 3D models are useful in the preoperative preparation and planning of operative strategy. The method is beneficial in cases of complex or rare CHDs where conventional imaging is difficult.

P-154
Assessment of tricuspid annular motion by speckle tracking in children with pulmonary arterial hypertension secondary to ventricular septal defect

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Introduction: Accurate evaluation of right ventricle (RV) function is fundamental in the management of pulmonary arterial hypertension (PAH) in children. Tissue motion annular displacement (TMAD) of the tricuspid annulus is an angle-independent method to assess annular motion. The aim of the study was to evaluate the utility of TMAD along with RV longitudinal strain, in assessment of RV function in children with severe pulmonary arterial hypertension secondary to ventricular septal defect (PAH-vsd).
Material and methods: We prospectively evaluated twelve children with PAH-vsd and twelve sex and age match controls. Beside conventional echocardiography, speckle tracking analysis of the right ventricule was performed using Q LAB 10.0 software. The following parameters were studied: tissue motion annular displacement (TMAD) of the tricuspid annulus (lateral, septal and midpoint), longitudinal strain of the RV (free wall and global), TAPSE (M-mode), right ventricular fractional area change, right ventricular myocardial performance index, left ventricular excentricity index and brain natriuretic peptide.
Results: TMAD of the tricuspid annulus (lateral, septal and midpoint) and RV longitudinal strain (free wall and global) were significantly lower in PAH-vsd children than in controls (p=0,0003, p<0,0001, p<0,0001 for TMAD, respectively p=0.0001, p=0.0001 for RV longitudinal strain). TMAD significantly correlated with all the studied parameters. TMAD showed the best correlation with the M-mode measured TAPSE, and with RV free wall and global longitudinal strain (r= 0,82, r= 0,58, r= 0,64, with a p< 0.05 for TMAD lat; r= 0,77, r= 0,64, r= 0,70, with a p< 0.05 for TMAD septal, respectively r= 0,83, r= 0,61, r= 0,67, with a p< 0.05 for TMAD midpoint).
Conclusion: Tricuspid Annular Motion assessment by Speckle Tracking (TMAD), along with longitudinal strain indices of the RV, is useful in noninvasive assessment of right ventricle performance in PAH-vsd children.

Key words: speckle-tracking echocardiography, tissue motion annular displacement, pulmonary arterial hypertension, ventricular septal defect, children.

P-155
Oxygen saturation lowering develops on the background of cardiac depression in Fontan patients

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Introduction: Fontan patients are likely to possess low oxygen saturation (SatO2), even if they do not own obvious right-to-left shunts, such as pulmonary arteriovenous fistula and fenestration from conduit to atrium. Such people sometimes complain of heart failure symptoms. We predicted degradation of cardio-pulmonary circulation would also provoke low SatO2.
Methods: The medical records of 382 Fontan patients were reviewed aged from 1 year to 41 years. They underwent cardiac catheterization between 2010 and 2018. We defined Low SatO2 as SatO2 90% or under (n=65). Cardio-pulmonary indexes were determined which were connected with Low SatO2 (n=86) by monovariate and multivariate analysis.
Results: Nine cardio-pulmonary factors were significantly different between Fontan patients with and without Low SatO2. Employing 283 patients who possessed no lost data for these 9 factors (Low SatO2: n=67), we investigated relevance indexes to Low SatO2. In mono-variate analysis, following factors were significantly related to low SatO2, such as end-diastolic ventricular volumes ≥ 205% (p=0.0030), end-systolic ventricular volumes ≥ 108% of Normal ventricular volumes on end-diastole (p=0.022), end-diastolic ventricular pressures ≥ 14 mmHg (p=0.020), end-systolic ventricular pressures ≤ 64 mmHg (p=0.035), pulmonary capillary-wedge pressures ≥ 13 mmHg(p<0.001), central venous pressures ≥ 17 mmHg(p=0.011), pulmonary artery indexes ≤ 123 mm2/m2 (p<0.001), pulmonary arteriovenous fistula (p=0.024), and fenestration from conduit to atrium (p<0.0001). After multivariate analysis, Low SatO2 was independently associated with following 5 factors: lowered ventricular pressures on end-systole (Odds ratio 7.8: p=0.035); elevated pressure of pulmonary capillary wedge (QR 5.5: p<0.001); narrow pulmonary arteries (OR 6.5: p=0.010); pulmonary arteriovenous fistula (OR 63.9: p<0.001); fenestration (OR 10.1: p<0.001). Explanatory coefficient was high (0.42) for low SatO2 by these 10 factors.
Conclusion: Our study showed that Low SatO2 was associated with cardio-pulmonary disorders other than apparent right-to-left shunts in Fontan patients. All aberrant values that we sought in this study were far outside, which indicated that Fontan circulation was going to collapse. If we detected Low SatO2 newly at a hospital visit, we should perform a variety of examination to suspect serious disturbance of cardio-pulmonary circulations.

P-156
Values of gamma-glutamyl transpeptidase are elevated in Fontan patients due to depressed cardiac functions as well as venostasis

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Introduction: Hepatic impairment in Fontan patients is considered to be provoked by liver congestion which is produced under high central-venous pressure. However, hepatic impairment does not exactly occur in Fontan patients with highly elevated pressure of central vein (CVP). In addition, we often find levels of gamma-glutamyl transpeptidase (GGT) highly elevated in Fontan patients whose cardiac functions seem to break down. We predicted hypo-cardiac functions would be major causes about hepatic damages in Fontan patients.
Methods: The medical records of 229 Fontan patients were reviewed aged from 2 tears to 46 years. They underwent cardiac catheterization between 2010 and 2017. We defined GGT 100 UI/L or over was GGT-elevation (n=50). Cardio-pulmonary indexes were determined which were connected with GGT-elevation in

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Fontan patients. We sought cardiac performances and pulmonary circulation factors related to GGT-elevation.
Results: We obtained following cardio-pulmonary performances related to GGT-elevation in Fontan patients: end-diastole ≥ 142% (p=0.0013); ventricular volumes on end-systole ≥ 72% (p=0.0013); ejection fraction of ventricle ≤ 38% (p=0.00014), cardiac output ≤ 1.7 L/min/m2 (p<0.0001), and moderate regurgitation of atrio-ventricular valve (p=0.011), CVP ≥ 15 mmHg (p=0.035), and pressures of pulmonary capillary wedge ≥ 9 mmHg (p=0.024). After multivariate analysis, GGT-elevation was independently associated with odds ratio of 11.1 for low cardiac output, 5.8 for decreased ventricular ejection fraction, 2.9 for expanded ventricular volume on end-diastol. Fontan patients with GGT-elevation took more amounts of carvedilol and enalapril than those without GGT-elevation. The values of NT-proBNP were higher in GGT-elevation group (962 vs. 216 pg/ml: p=0.00029). The rate of patients were higher in GGT-elevation group who underwent catheterization owing to having some sort of symptoms or abnormal findings (32% vs. 15%: p=0.0054).
Conclusion: Our study showed that GGT-elevation in Fontan patients was not independently associated with CVP elevation but depression of cardio-functions. Cardiac hypo-functions would injure liver by hepatic hypo-perfusion. Absolutely, venostasis, which would diminish the liver, is happening in Fontan patients with CVP elevation. However, CVP elevation would be induced by high afterloads against pulmonary arteries, which was provoked cardiac hypo-function. We should tighten up medical treatment strongly for heart failure to preserve hepatic functions.

P-157
Imaging parameters predictive for exercise capacity in patients after the arterial switch operation

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Background: The arterial switch operation (ASO) for transposition of the great arteries has excellent survival, but a substantial number of patients suffers from a reduced exercise capacity. The goal of this study was to identify imaging parameters predictive for a reduced exercise capacity in patients after ASO.
Methods: A retrospective analysis was performed of ASO patients who underwent cardiopulmonary exercise testing (CPET) between 2007 and 2017. Reduced exercise performance was defined as a reduced workload peak (Wpeak) with Z-score <-2 or a peak oxygen uptake indexed for weight (VO2peak/kg) with Z-score <-2. Data on echocardiography and cardiac magnetic resonance (CMR) performed within 1 year of the CPET were collected for comparison.
Results: A total of 81 ASO patients (age 17±7 years) were included. Reduced exercise performance was found in 17 patients (21%) as expressed by a reduced Wpeak and 15 patients (19%) with a reduced VO2peak/kg. Left pulmonary artery stroke volume by CMR, and main pulmonary artery gradient and tricuspid regurgitation gradient by echocardiography were found to be predictive of reduced Wpeak (p=0.008; p=0.009; p=0.026, respectively). The main pulmonary artery gradient and tricuspid regurgitation gradient by echocardiography were found to be predictive of reduced VO2peak/kg (p=0.031; p=0.035, respectively).
Conclusion: This study demonstrates that ASO patients frequently experience reduced exercise capacity. Imaging parameters of main pulmonary artery and pulmonary artery branch stenosis and associated increased right ventricle pressure were predictive for reduced exercise capacity, and are therefore key during serial follow-up of ASO patients.

P-158
Evaluation of pentraxin 3 level and cardiac functions in psoriatic children

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Introduction: Psoriasis is a chronic inflammatory disorder affecting the skin, nails, and joints. Its prevelance has been estimated to be at 1% to 3% with lifetime in population. This study was designed to examine the association between serum pentraxin 3 (PTX3) and cardiovascular function in psoriatic children.
Methods: 33 children who were diagnosed with psoriasis, and 29 healthy children, between 4 and 18 years of age, were included in the study. Both patient and control group was evaluated by the pulsed wave tissue Doppler echocardiography (TDI), as well as with conventional Doppler echocardiography (CDE). PTX3 values of the groups were evaluated.
Results: There was no difference between cases and controls for age (9.67±3.72, 9.60±2.84 years, p=0.916, respectively). In evaluation of LV (left ventricle) CDE; A wave, isovolumic relaxation time (IVRT) and myocardial performance index (MPI) were significantly higher in study group (p<0.05). Ejection time(ET) was significantly lower in study group compared to control group (p<0.05). In evaluation of LV TDI; Deceleration time (DT'), IVRT', E/E' and MPI' were found to be significantly higher in study group (p<0.05). In addition to, E', E'/A' and ET' were siginificantly lower in study group. PTX3 level was significantly higher in the study group compared to the control group (p=0.009)(Table I). However, no correlation was found between PTX3 level and cardiovascular parameters.
Conclusion: Both doppler echocardiography and PTX3 may be useful tools for the screening of CV risk in these patients. Psoriasis itself may be an independent risk factor for cardiac dysfunction in pediatric population.

Table I. CDE, TDI parameters and PTX3 values of groups

Parameter	Study (n = 33)	Control (n = 29)	P
E (cm/s)	82.1 ± 11.6	80.4 ± 11.7	.569
A (cm/s)	49.9 ± 13.1	43.2 ± 9.5	.026
E/A	1.73 ± 0.46	1.93 ± 0.45	.095
S (cm/s)	79.6 ± 16.26	85.3 ± 10.8	.117
DT (ms)	89.7 ± 19.5	84.5 ± 12.9	.234
ET (ms)	241.1 ± 19.4	257.5 ± 17.0	.001
IVRT (ms)	71.2 ± 16.5	58.9 ± 12.4	.002
IVCT (ms)	66.9 ± 16.4	68.0 ± 13.0	.768
MPI	0.57 ± 0.10	0.49 ± 0.09	.002
E' (cm/s)	15.33 ± 3.41	17.55 ± 2.64	.006
A' (cm/s)	7.21 ± 1.89	6.72 ± 1.19	.239
E'/A'	2.21 ± 0.62	2.67 ± 0.52	.003
E/E'	5.71 ± 2.21	4.67 ± 0.94	.023
S' (cm/s)	9.81 ± 2.44	9.06 ± 1.22	.141
DT' (ms)	74.90 ± 12.59	64.03 ± 11.62	.001
ET' (ms)	256.5 ± 29.3	271.7 ± 20.8	.024
IVRT' (ms)	60.30 ± 10.36	52.65 ± 11.19	.007
IVCT' (ms)	66.51 ± 15.33	61.89 ± 9.23	.163
MPI'	0.50 ± 0.11	0.42 ± 0.07	.003
PTX3 (ng/ml)	5.89 ± 5.00	3.10 ± 2.61	.009

E, early diastolic myocardial velocity; A, late diastolic myocardial velocity; S, systolic myocardial velocity; DT, deceleration time; IVRT, isovolumic relaxation time; IVCT, isovolumic contraction time; ET, ejection time;
MPI, myocardial performance index; PTX3, pentraxin 3.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-159

Pulmonary Artery Optical Coherence Tomography in Pediatric cases of Pulmonary Arterial Hypertension

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Introduction: Optical Coherence Tomography (OCT) is a high-resolution (10microns) intravascular imaging technique using near infra-red light. Widely used to image coronary arteries, pulmonary artery (PA) imaging is less well described. We performed OCT on the pulmonary arteries of children with pulmonary arterial hypertension and controls with the hypothesis that alterations within the vessel wall of the pulmonary arteries could be seen in association with pulmonary arterial hypertension.
Methods and Results: Pulmonary artery OCT was performed in six patients plus ten controls. Subjects were from 2-11 years of age (4 female). Diagnosis; idiopathic pulmonary hypertension (PHT) 2; late repair left to right shunt 2, repair Trisomy 21 with associated shunt lesion n=2. Medication at time of study; Nil n=1, phosphodiesterase inhibitor/endothelin inhibitor n=2, triple therapy with Treprostinil n=2. Pulmonary Vascular resistance (on therapy) 3-10 Woods units; nil therapy 12 Woods units. Control patients were age and weight matched undergoing either diagnostic or electrophysiology study. Serial measurements were made along the length of the vessel using digital planimetry to calculate wall thickness (mm) and wall:vessel cross sectional area (CSA) ratio for each case. Median pulmonary artery wall thickness for the pulmonary arterial hypertension patients was 0.18mm (IQR 0.17-0.21mm) and for controls was 0.11mm (IQR 0.10-0.12mm) (p 0.002). Median pulmonary artery wall:vessel CSA ratio for the pulmonary arterial hypertension patients was 0.19 (IQR 0.19-0.22) and for controls was 0.13 (IQR 0.11-0.15) (p = 0.002). As there was a significant difference between our pulmonary arterial hypertension and control groups with regard age and weight, we made an additional comparison with a younger control group. A statistically significant difference persisted for wall thickness and wall:vessel cross sectional area ratio (p=0.025). There were no complications.
Conclusion: Pulmonary artery OCT is feasible in children and identifies increased wall thickness in pulmonary arterial hypertension. PA OCT can be used to recognise reverse PA wall remodelling in response to treatment. Further studies are required to assess if OCT can help distinguish between causes of pulmonary arterial hypertension in children and degree of vessel fibrosis.

P-160

Liver and spleen assessment in post-Fontan pediatric patients

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Introduction: Fontan-associated liver disease (FALD) is one of the consequences of Fontan hemodynamics affecting long-term prognosis. Magnetic resonance elastography (MRE), a relatively new imaging technique measuring tissue stiffness, provides estimation of liver fibrosis and congestion. Spleen stiffness is considered to be dependent on the degree of liver fibrosis and can be also evaluated by MRE. Moreover, its degree should correlate with the risk of hypersplenism and esophageal varices bleeding. To the best of our knowledge, we describe the first reported data of spleen involvement in post-Fontan pediatric patients.
Methods and material: Detailed follow up (both cardiac and multi-organ evaluation) was planned to assess long term consequences of Fontan hemodynamics in a group of 30 pediatric patients. Hepatic and spleen screening involved MRE, abdominal ultrasound and laboratory studies. Present study include a group of 11 patients with hepatic and spleen screening data available. We evaluated the presence of liver and spleen pathology and analysed their importance for post Fontan patients.
Results: There were patients 5 to 12 years post-completion, 9 of them with systemic left ventricle. All patients had elevated spleen and liver stiffness values, the first being higher in all but two patients. Mean spleen stiffness was 5,8kPa with maximum 10kPa comparing to mean liver stiffness 4,34kPa with maximum 5,2kPa (reference range for liver stiffness is 1.54 to 2.87 kPa, for spleen slightly larger but not yet established). Splenomegaly and/or hypersplenism was detected in 4 patients, esophageal varices in 1 patient. Correlation between these findings and severity of spleen or liver stiffness was not confirmed. Moreover, 1 patient had splenic haemangioma, but the relation of this finding to Fontan circulation is unknown.
Conclusions: Magnetic resonance elastography is promising in assessment of liver as well as spleen in Fontan circulation. Presence of advanced spleen pathology suggests to implement screening for hypersplenism and esophageal varices in routine long-term follow up of Fontan patients. As this very first reported results are potentially important, they should be confirmed in future studies.

P-161

Product of Peak Systolic Wall Stress and Heart Rate Detects Preclinical Cardiomyopathy in Duchenne Muscular Dystrophy

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Introduction: Cardiomyopathy is a common late complication in Duchenne Muscular Dystrophy (DMD), but its onset is insidious and not clearly defined. We proposed "wall stress index (WSI)" as a marker of total left ventricular (LV) workload and tested whether it can detect preclinical myopathic changes in DMD.
Methods: Peak systolic wall stress (PSWS) was calculated in M-mode echocardiography with simultaneous measurement of systolic blood pressure (SBP). WSI was defined as PSWS x heart rate (HR). We measured WSI and LV mass index (LVMI) in normal controls, DMD with normal LV fractional shortening (FS > 30%) (DMD-A), and DMD with decreased LVSF (< 30%) (DMD-B). The data were shown as mean ± standard deviation.
Results: Total 36 normal controls and 47 DMD patients (83 DMD studies; 36 patients had two studies) were investigated. DMD-A was divided into two groups at the age of 10 years. HR was significantly higher in DMD groups than in controls. Despite comparable FS, WSI was significantly higher in DMD-A, even < 10 years, than in controls. WSI became even higher as FS further decreased (DMD-B). All DMD-B patients were treated with an angiotensin II-converting enzyme inhibitor and/or a β-blocker. In a longitudinal study with 33 DMD patients over 1 to 8 (4.0 ± 2.0) years, WSI showed a significant increase (p = 0.01) with a decline in FS. LVMI remained mostly within normal limits but significantly increased with age (p = 0.021).
Conclusions: Our data demonstrated that 1) WSI was significantly higher in young DMD than in controls despite comparable FS and that 2) WSI further increased as FS declined with age. The increase of WSI preceded the decrease in FS, suggesting its diagnostic value in detecting preclinical cardiomyopathy in DMD.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

	Controls	DMD-A (< 10 yrs)	DMD-A (> 10 yrs)	DMD-B
Number	36	23	28	32
Age (yrs)	13.2 ± 2.9	7.1 ± 1.7	14.1 ± 3.2	16.4 ± 3.7
HR	70 ± 16	92 ± 13*	91 ± 13*	88 ± 13*
SBP (mmHg)	103 ± 13	92 ± 13*	105 ± 12	101 ± 11
FS (%)	36.8 ± 3.5	35.9 ± 3.0	34.8 ± 4.3	23.0 ± 4.4*
WSI	295 ± 68	444 ± 123*	483 ± 132*	686 ± 213*†‡
LVMI	29.8 ± 8.8	35.0 ± 5.9	32.6 ± 5.9	34.2 ± 12.2

$p < 0.05$ compared with control*, DMD-A (< 10 yrs)†, and DMD-A (≥ 10 yrs)‡

Longitudinal Follow Up of 33 DMD Patients (4.1 ± 2.0 years)

	Before	After	p value
Age (yrs)	11.7 ± 3.6	15.8 ± 4.2	
%FS	32.3 ± 6.0	28 ± 7.3	$< 10^{-4}$
WSI	507 ± 163	600 ± 206	0.01
LVMI	31.6 ± 9.5	35.0 ± 10.5	0.021

P-162
Normal myocardial T1 values in children using a saturation and an inversion recovery sequence

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Objectives: T1 mapping by cardiac magnetic resonance (CMR) allows detection of diffuse myocardial processes such as fibrosis, edema, storage disorders, or hemochromatosis. MOLLI is a widely used inversion recovery sequence that measures T1*, which is then mathematically converted into T1 values. Smart1Map is a new saturation recovery sequence that measures T1 directly. Our aim was to establish normal values for Smart1Map in children and to compare them to MOLLI.
Methods: Twenty-nine children between 8 and 18 years of age (14 males) without evidence of cardiovascular diseases prospectively underwent CMR on a 1.5T GE scanner. Ventricular volumes and function were assessed by SSFP cine, and T1 values assessed with MOLLI and Smart1Map sequences. T1 values of myocardium and blood were determined from the resultant maps in three short-axis slices (basal, mid-ventricular, apical) using QMap (MEDIS, Leiden, NL). A region of interest was defined in the interventricular septum of each slice.
Results: In all slices, T1 values were higher by Smart1Map than by MOLLI (Table 1). Significant differences in myocardial and blood T1 values were observed between different slices by MOLLI (myocardium $p<0.001$; blood $p<0.05$), but not by Smart1Map (myocardium $p=0.077$; blood $p=0.59$). Myocardial T1 did not correlate with heart rate when assessed by either method. After careful exclusion of artifacts, no differences were found between septal and total myocardial T1 values at each slice using either method.

Table 1: Myocardial T1 values by Smart1Map versus MOLLI

	T1 Smart1Map [ms]	T1 MOLLI [ms]	
Basal myocardium	1197 +/- 63	996 +/- 35	p<0.001
Mid-ventricular myocardium	1196 +/- 63	1019 +/- 41	p<0.001
Apical myocardium	1223 +/- 60	1053 ± 48	p<0.001

Conclusions: We established pediatric normal values for native T1 mapping using the Smart1Map sequence and compared the results to T1 mapping with MOLLI. Smart1Map showed more robust T1 values among different myocardial slices than MOLLI. Septal values can be used to represent the whole myocardium in cases with suboptimal image quality and diffuse pathology.

P-163
Transcatheter closure of ventricular septal defects with the off-label occlusion devices

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Background: Transcatheter closure or ventricular septal defects (VSDs) is increasing by the time. The most frustrating problem with closure of VSD is the complete heart block. So searching for different devices is ongoing and unfortunately there were no optimum device for this procedure yet. We present to evaluate the safety and efficacy of the off-label devices for the transcatheter closure of VSDs.
Method: Patients who underwent transcatheter VSD closure with off-label devices between 2014 and 2018 were reviewed retrospectively.
Results: VSD was closed in 30/31(96%) of the patients. Seventeen of the patients were boys and 13 were girls. The median age and weights of the patients were 4.8 years(1.4-23.6 years) and 17 kg (9-70kg) respectively. The localization of the VSD was perimembranous in 27 and muscular in 3 and 25/27 patient has ventricular septal aneurism. VSDs were closed with Amplatzer Duct Occluder (ADO)-I (n=15), Amplatzer Vascular Plug (AVP)–II (n=14), ADO-II (n=1). The median age was 3.1 years in the ADO patients, and 6.7 years in the AVP-II patients. The narrowest median VSD diameter on the angiogram was 5 mm (3-10.2mm). The median fluoroscopy time was 14.6 min (5.5-35.6 min). Patent ductus arteriosus (PDA) was also present in two patients was closed during the same session. Also the left ventricular-right atrial shunt decreased or disappeared in five patients after the procedure. In one patient procedure was unsuccessful due to the ventricular tachycardia when the arteriovenous loop was performed. No patient had complete atrioventricular block during the 20 months (1-55 months) follow-up period. Two patients had left bundle branch block and they are still under follow-up. Three patients were followed due to the mild residual shunt across the device, and 1 to 10 months after the procedure all the residual leak was disappeared.
Conclusion: Some devices can be used off-label for transcatheter VSD closure in selected patients. ADO-1, ADO-2 and AVP-2 seems an effective and safe treatment option in selected patients. Because of the fact that these devices are more flexible and softer than the other devices and also no disks on the right ventricle side may lead to less rhythm problems.

P-164
Use of Amplatzer vascular plug II device to occlude different types of patent ductus arteriosus in pediatric patients

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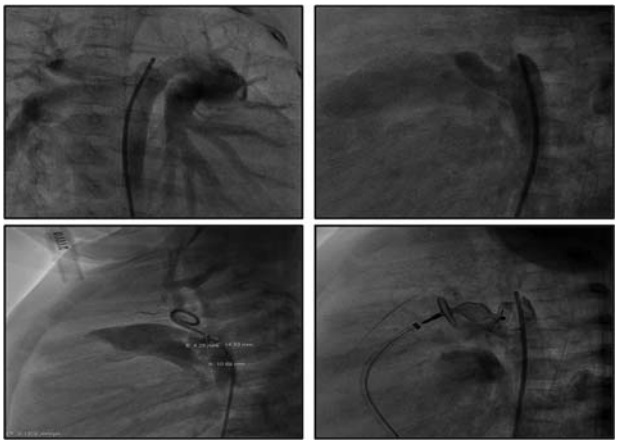
https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



Objectives: To evaluate the outcome of the Amplatzer Vascular Plug II (AVP II) for closure of patent ductus arteriosus (PDA) in children.
Methods: All patients undergoing transcatheter closure of PDA with AVP-II from June 2014 to November 2018 were retrospectively evaluated. Clinical, angiographic, and echocardiographic data were analyzed.
Results: Eighty-four procedures were performed. Amplatzer Vascular Plug II was used in 81 patients. Median age was 24 months (3 months–16.8 years) with median weight 13 kg (4.2–72). The morphological PDA classification was Type A in 37(45.6%), Type B in 1(1.3%), Type C in 17(21%), Type D in 1(1.3%) and Type E in 25(30.8%). The median minimum, maximum PDA diameter and length were 3.1 mm, 8.4 mm and 12 mm, respectively. The implanted AVP II device sizes were: 6 mm in 5/81 (6%), 8 mm in 43/81(53%), 10 mm in 25/81 (31%), 12 mm in 1/81(1%), 14 mm in 5/81(6%) and 16 mm in 2/81(3%) procedures. The implanted device was mean of 2.8(1.7-6.8) times the ductus narrowest diameter and mean of 1(0.4-3.2) times the ductus largest diameter in successful procedures. The median procedure and fluoroscopy time was 25 minutes and 4.6 minutes. In 12(15%) patients closure was performed from the arterial side and in 69(85%) venous access was used. All procedures except one were successful; among successful procedures 100% 'in-lab' and 100% closure on post-procedural echocardiogram was achieved. In two (2.5%) patients, the device was embolised; one patient underwent surgery and in the other patient, 8mm device was retrieved and a 10 mm device was implanted on the next day. Left pulmonary artery stenosis was observed in three (3.5%) patients; one patient underwent surgery and the degree of pulmonary stenosis was mild in two. Aortic obstruction observed in only one patient with 15 mmHg gradient.
Conclusions: The AVP II seems to be an effective and safe device for PDA closure in children. It is particularly useful in type C and E ductus and in small infants where it eliminated the risk of device related aortic obstruction.

P-165

Novel high-flow microcatheter and large volume dense detachable coils for closing hard to access aberrant arterial vessels in children

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Introduction: Accessing and closing arterial vessels in infants and young children can be challenging due to their acute angle of origin or long tortuous course. Standard catheters are often too thick and stiff for intubation and especially placement of occlusion devices in peripheral position. Furthermore, often only incomplete occlusion of large vessels is achieved.
Methods: The Lantern Catheter (Penumbra Inc., USA) is a 2.6F high-flow microcatheter that allows for accessing tortuous vessels and placement of dense packing coils (Ruby Coils) to completely occlude aberrant arterial vessels. We report our experience in transcatheter occlusion of hard to access major aortopulmonary collateral arteries (MAPCA) and large, high-flow pulmonary sequestration arteries in three patients using the Lantern Catheter and Ruby Coils.
Results: Patient 1 is a four-year-old male with uncorrected Tetralogy of Fallot and pulmonary sequestration (PS) of the right lower lobe whose hard to access supplying artery arises from the thoracic descending aorta in an acute angle.
Patient 2 is a 7-year-old male with right lower lobe PS whose large supplying artery arises from the abdominal aorta cranial to the coeliac duct and draining after a tortuous course into the right pulmonary veins.
Patient 3 is five-year-old girl with double inlet left ventricle, s/p Fontan-completion with MAPCAs arising from the right subclavian and costocervical artery.
Despite unsuccessful occlusion during previous catheterizations in two of the patients we were able to close the aberrant vessels successfully in all of them.
Conclusion: The Lantern Microcatheter and dense packing Ruby Coils are very valuable in closing PS arteries and MAPCAs. This system may become an alternative in infants and young children for closing hard to access, tortuous vessels and large arteries with high blood flow.

P-166

Transcatheter Embolization Of Coronary Artery Fistulas in Pediatric Patients: Clinical, Angiographic Findings And Long-Term Follow-Up Results Of A Single Center

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Introduction: Coronary artery fistula (CAF) is a direct connection between a coronary artery and one of the cardiac chambers or great vessels. CAFs account for 50% of all congenital coronary anomalies, presenting in 0.002% of general population. Clinical presentation varies depending on size of fistula, age, presence of myocardial ischemia. Most patients are asymptomatic during childhood. Closure of CAFs are recommended even in asymptomatic patients to prevent fistula related complications ('steal' from the adjacent myocardium, thrombosis/embolism, heart failure, atrial fibrillation, rupture, endocarditis/endarteritis, arrhythmias). We aimed to evaluate clinical,angiographic,surgical findings, and long term follow-up results of CAF patients diagnosed between 2000-2018. Methods: Clinical,catheterization,surgical data of patients diagnosed as CAF were retrospectively analyzed. Results: CAFs were diagnosed in 38 patients(mean age:6.5±5.3 years (3 days-17years),weight:25±19kgs(3.5-75),24 male,14 female). 58% of patients presented with murmur, in the remaining CAFs were incidentally diagnosed by echocardiography(16%), or with an associated defect(18%). Associated anomalies were DORV/VSD(1), bicuspid aortic valve/severe valvular AS(1),

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

operated subaortik ridge/PDA(1), bicuspid aortic valve(1), AVSD (2), large ASD(1).Diagnosis were confirmed by catheterization in (35/38) or by computed tomography angiography(3/38). 34/38 patients(90%) had single, 4/38(10%) had multiple CAFs. Origin of fistula were left coronary artery(LCA)(17), right coronary(10), left anterior descending (LAD)(9), circumflex artery(2) and they terminated in pulmonary artery(13),right ventricle(13), left atrium(5), left ventricle(2),right atrium(4),coronary sinus(1). In 24/38 patients(63%),proximal coronary artery from which CAFs arise had dilatation. Transcatheter embolization were performed in 16/38 patients(42%)(mean age:4.6.±5.0 years(14 days-16.5years), weight:20±19kgs(4-75),successful in 14/16(88%).Embolization were not performed in 22/38 patients(58%) due to small fistula size and followed clinically. Occlusion devices were Amplatzer Vascular Plugs(6) coils(6), Amplatzer Duct Occluder II Additional size device (3), glue (cyanoacrylate)(1). Mean procedure time:71±52min (40-220),fluoroscopy time:23±13min(2-60). No complications were observed. Surgery were necessary in 5/38 patients(13%), for unsuccessful embolization(2) or during congenital heart disease surgery (3). Mean follow-up period was 4.3±4.0 years(1 month-16 years). During the follow-up, no patients had recurrence of fistula. 2/38 patients(0.05%) had spontaneous occlusion. Control angiography were performed in 12/16 patients after successful embolization, 3/12 had remodelling, regression of the size of the involved coronary artery. Conclusions: Transcatheter embolization of CAFs is an effective and safe procedure in pediatric patients. Follow-up is necessary for early diagnosis of recurrence of fistula.

P-167
Aortic balloon valvuloplasty and mid-term results in newborns; single center experience

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Objective: Aortic balloon valvuloplasty (ABV) has become the first-line treatment for critical aortic valve stenosis. We aimed to evaluate the short- and mid-term results of patients who underwent ABV during neonatal period, the factors affecting the success and complications.
Methods: We retrospectively examined 65 patients who underwent ABV during the neonatal period between 1998 and 2017. Cardiac catheterization reports, echocardiographic information, and angiographic views were reviewed for the factors affecting procedural success and aortic regurgitation (AR), complications, the need for repeat ABV or aortic valve replacement.
Results: Forty-five (69.2%) of the patients were male and mean follow-up was 6.2±4.9 years (6 months-19 years). The mean age at the first ABV was 14.5±10.6 days (1-30) and body weight was 3.25±0.6 kg (1.5-4.8). The peak systolic gradient at pre-valvuloplasty cardiac catheterization was 73.3±22.7 mmHg (30-142 mmHg), and it decreased to 29.2±12.2 mmHg (5-55 mmHg) after the procedure. Valvuloplasty was successful in 59 (90.7%) patients. Four patients underwent surgical valvatomy. There was no more than mild AR in any patient before valvuloplasty. In the acute phase after valvuloplasty, 30 patients had mild, 15 had moderate and two had severe AR. There was a significant increase in the degree of AR related to valvuloplasty ($p<0.05$). The most important complication of ABV was increased AR (26.2%). Another important complication was femoral artery occlusion detected early after valvuloplasty (61.6%). There was no serious complication or death in the acute phase. Six patients died in the first year, four of them had Shone complex. Six patients had aortic valve replacement due to severe AR.
Conclusion: In newborns with valvular aortic stenosis, balloon valvuloplasty has become the first choice in many centers due to its high success rate, low mortality and morbidity. Aortic regurgitation and femoral artery occlusion were the most important complications. Although reintervention for residual or recurrent aortic valve stenosis is common during the first year after valvuloplasty, these patients are able to reach advanced ages without the need for surgical intervention. Surgical valvotomy is a good alternative treatment for a small number of patients in whom valvuloplasty fails.

P-168
Transcatheter correction of partial anomalous pulmonary venous connection

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Introduction: Partial anomalous pulmonary venous Drainage is an extremely rare congenital Defect. Patients with a partial anomalous pulmonary venous Drainage, if symptomatic or showing significant shunting, are generally treated with surgery.
Method: A Fifteen-year-old adolescent was admitted to our Department suffering from reduced exercise tolerance and Dyspnea at exercise. The echocardiogram revealed a dilated right ventricle without atrial septal defect. The patient underwent cardiac MRI, which showed a left pulmonary vein draining to the hepatic veins with minimal drainage to left atrium. A cardiac catheterization was performed: Angiography revealed A significant left-to-right shunt via the anomalous venous connection from the left pulmonary vein via a vertical vein into liver veins and a minimal Shunt from the single left pulmonary vein into left atrium (dual drainage). After occlusion of the vertical vein with a 30 mm Sizing balloon angiography oft he left pulmonary artery revealed drainage of the left pulmonary vein to left atrium without any obstruction. A 16 mm Amplatzer Vascular Plug II (AVP II) was used to occlude the vertical vein.
Result: After deploying of the 16 mm AVP II angiography showed an unobstructed blood drainage to the left atrium, no residual leak over the vertical vein.
Conclusion: Transcatheter therapy In partially abnormal pulmonary venous return with dual drainage is feasible and safe. It offers a good alternative to surgery in selected cases.

P-169
Initial outcome of perventricular device closure of ventricular septal defects in children without cardiopulmonary bypass

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Introduction: Ventricular septal defects (VSDs) are estimated to account for 20% to 30% of all congenital cardiac malformations and are the commonest CHD excluding Bicuspid aortic valve. Open surgical and percutaneous transcatheter approach are the two well-known methods with their advantages and disadvantages. Off late a new hybrid approach, perventricular closure of

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

VSDs is gaining importance given the benefits it has over both these procedures.
Methods: We aimed to analyse the success and incidence of acute complications after perventricular device closure of ventricular septal defect (VSD) without cardiopulmonary bypass in children. Children aged less than 18yrs, with isolated VSD, who underwent perventricular device closure of VSD without cardiopulmonary bypass, from September 2017 to November 2018 were included. Inclusion criteria for the study were congestive cardiac failure refractory to medical management, and children who had percutaneous VSD device failure (if deemed suitable after Transthoracic echocardiography, and after on table Transoesophageal echocardiography assessment for device suitability). Children with either of the following criteria were excluded: VSDs with significant aortic prolapse, VSDs with inlet extension and associated lesions requiring open heart surgery.
Results: A total of 31 children (M: F= 0.93) were included in the study. The median age of the study population was 9 months (IQR 75th, 25th: 25,6). Median weight of the study group was 6.7kg (IQR 75th, 25th: 8.5,5.3). Muscular VSD was the commonest (84%). Mean duration of the procedure was 90±26 min. Median post OP ventilation time was 12 hours (IQR 75,25: 18.5,8.7) and median hospital stay 7 days (IQR 75,25:8,5). There were 3 (9.6%) failures (new onset significant Tricuspid regurgitation, device induced haemolysis and device embolisation to Left pulmonary artery). Post-procedure, new trivial TR and new trivial AR which are haemodynamically not significant were seen in 42% and 3.2% of the cases, respectively. Post-procedure transient RBBB was observed in 1 child.
Conclusion: Perventricular approach for device closure of VSD is an effective alternative to traditional VSD closures. It has minimal complications with a good success rate.

P-170
Dealing with an unroofed coronary sinus in a delicate clinical condition
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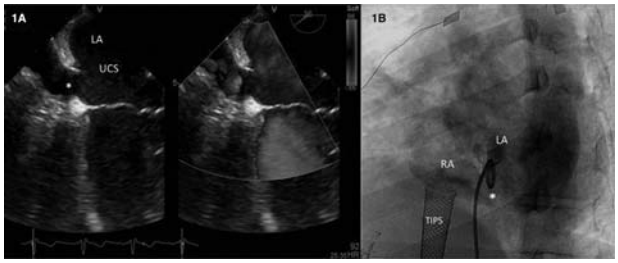
Introduction: Unroofed coronary sinus is an uncommon type of atrial septal defect in which a communication occurs between the coronary sinus and the left atrium as a consequence of an absent or deficient coronary sinus septum.
The diagnosis may be often overlooked because of the lack of specific clinical features and the difficulty to define the exact anatomy by transthoracic echocardiography since the defect is not always oval and the coronary sinus is not often a perfect cylinder.
Methods and Results: We describe a 17 year-old girl with hepatic failure that was assessed by a pediatric cardiology team before a liver transplant. She was affected of a portal cavernomatosis initially treated with a transjugular intrahepatic portosystemic shunt (TIPS) due to portal hypertension. Transthoracic echocardiography showed a right ventricular dilatation. Also, an unroofed coronary sinus (UCS) was suspected. No persistent left superior vena cava was observed. Magnetic resonance imaging confirmed a complete absence of the roof of the coronary sinus with an enlarged ostium (asterisk in the figure), and a Qp/Qs ratio of 1.9. In a multidisciplinary meeting a percutaneous approach to close the coronary sinus atrial septal defect type was decided given the delicate condition of the patient. Transesophageal echocardiography helped to define anatomy (Figure 1A) and to guide device position.



The right femoral vein was cannulated and an angiogram from the left atrium demonstrated the coronary sinus atrial septal defect (Figure 1B). A Figulla Flex II ASD occluderTM 27mm was successfully deployed. There was no residual shunt or atrioventricular valves interference, and the patient remained on sinus rhythm at all times. The device stopped the left to right transatrial shunt, leaving the unroofed coronary sinus draining to the left atrium. The right sided chambers dilatation normalized progressively, and six months later the device remains stable in a normal position.
Conclusions: Although surgery is still the mainstay for unroofed coronary sinus closure, transcatheter-based management is quickly evolving and it may represent a valid, safe and less invasive alternative to conventional heart surgery.

P-171
Balloon angioplasty of aortic coarctation in critically-ill new-borns using axillary artery access
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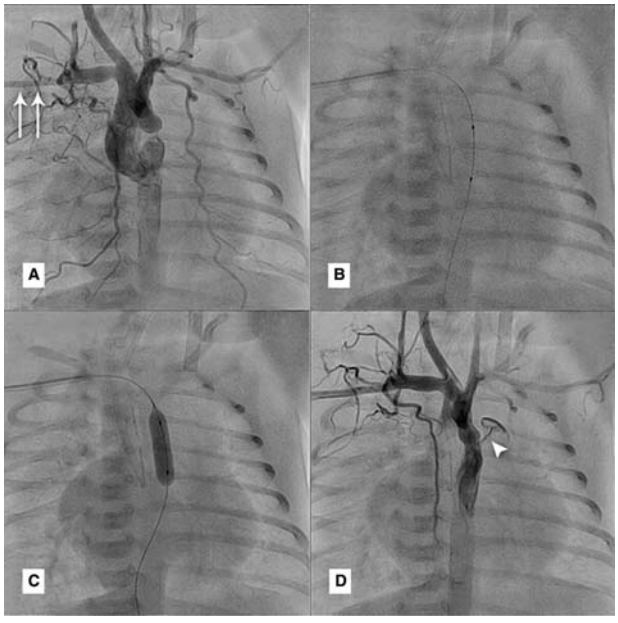
Introduction: Standard treatment for new-borns with native aortic coarctation is surgery. In selected cases balloon angioplasty may be performed, to stabilize neonates unsuitable for immediate surgery. The aim of this paper is to describe our experience in percutaneous treatment of aortic coarctation via axillary artery access, an alterative option to the traditional femoral and carotid artery routes.



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

	Sex/Age	Weight at procedure (kg)	Aortic arch hypoplasia	Concomitant CHD
C 1	F/2 mo	3.6	No	None
C 2	F/1 mo	2.3	No	BAV
C 3	F/4 mo	4.7	No	None
C 4	F/10 d	2.9	No	VSD+MS
C 5	M/4 mo	5.6	Yes	Bov Arch
C 6	M/13 mo	5.5	Yes	BAV
C 7	M/2 d	2.4	No	None
C 8	F/1 mo	1.8	No	None

	FUP (y)	Late complications	Further procedures
C 1	5	None	None
C 2	5	None	Aortic valvuloplasty (2 mo)
C 3	5	None	None
C 4	4.6	None	None
C 5	3.5	None	None
C 6	2.6	Re-CoA	Patch aortoplasty (2 m)
C 7	0.8	Re-CoA; Local aneurysm	Patch aortoplasty (2 m)
C 8	0.6	None	None

Case series: Ten consecutive patients were treated with urgent balloon angioplasty for aortic coarctation between 2012 and 2018. In eight of them the procedure was performed via axillary artery access. Pre-procedural data are summarized in Table 1. They were all cases of native aortic coarctation, with the exception of a patient already treated with patch aortoplasty for a hypoplastic aortic arch who presented with recurrent stenosis at repair site (case 5). All but this latter patient were critically ill infants, with clinical signs of low cardiac output, left ventricular dilation and dysfunction and lactic acidosis. Femoral pulses were non-palpable. Patients were therefore scheduled for urgent balloon angioplasty.In every patient right axillary artery was accessed and cannulated with a 4-F sheath. A 5-20-65 mm Tyshak-Mini balloon was advanced over a 0.014" floppy guide-wire across coarctation and inflated twice. Angiographic and pressure gradient control demonstrated effective angioplasty in five patients. In case 4 two additional inflations were carried out with balloons of progressive greater diameter. Haemostasis was always effectively achieved with digital compression at the puncture site for 60 minutes. A brief episode of bradicardia, immediately resolved with atropine, was the only minor complication reported (case 6). All patients are alive. Follow-up data are summarized in Table 2.
Conclusion: Axillary artery access may be considered a feasible, safe and effective alternative approach to the traditional artery access routes, even in smaller critical newborns with concomitant low cardiac output, as it displays some technical advantages. Axillary pulse is easier to feel in smaller patients, especially in premature new-borns and in presence of critical aortic coarctation, when femoral pulses are not palpable. Axillary artery is not an end-artery and thus, when cannulated, arm perfusion is still guaranteed by the second intercostal artery and the acromial artery.

P-172
Midterm experience with a novel PTFE-tube covered cobalt-chromium stent in pediatric and adult patient with coarctation of the aorta

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Objective: Treatment of coarctation of the aorta (CoA) by either ballooning or stent implantation is a recommended treatment form in children and adults. It may include subatretic, aneurysmatic, surgical or interventional pre-treated aorta, where the use of a covered stent is favorable in order to prevent dissection, aneurysm or rupture.
Methods: We report our midterm experience with the use of the novel ePTFE-tube covered cobalt-chromium stent (BeGraft, Bentley, Germany) for aortic stentgraft implantation during 01/2017 until 08/2018.
Results: Seventeen patients were included in this study with a median age of 17.8 (3.8–49) years. Ten (58%) out of these patients were below 18 years of age and seven (41%) had a pretreated CoA (by either surgery or intervention). One patient received a bypass (Art. carotis to subclavia) 3 month prior intervention. The median weight was 55.8 (18-101) kg. Pre-catheterization imaging was performed with either MRI or CT and guidance of the catheter procedure by image fusion software was used with 3D overlay. All stent were successfully implanted: 4x 12/29, 2x 14/29, 1x 14/59, 1x 16/38, 2x 16/48, 1x 20/48, 2x 22/48, 3x 24/48mm through a 9-14Fr. sheath. Rapid pacing was performed in two, post dilatation in 7 patients with high pressure balloon (12-22mm) in order to achieve a post gradient < 5mmHg. Dysfunction (aneurysm, residual gradient, dislocation) was excluded during follow-up by using echocardiography, X-ray, RR-measurement or imaging (CT/MRI) at 3, 6 and 12 month. The median event free follow-up time was 0.72 (0.1–1.64) years.
Conclusion: This is the first report on a larger cohort with aortic stent-graft implantation in native or pre-treated CoA with the use of a pre-mounted cobalt-chromium stent-graft covered with micro-porous ePTFE tubing, since the stent received CE-mark approval in 12/2016. The pre-mounted stent showed a good radial force, reliable coverage, adequate adaptation to complex anatomy and promising function during midterm follow-up time in children and adult patients. Additional data is still necessary in order to demonstrate efficacy and long term performance of this stent.

P-173
Amplatzer Vascular Plug II: The ideal device for closure of type E and C ducts

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Introduction: The indications for patent ductus arteriosus (PDA) closure for preterm babies in the neonatal period have been decreasing in some centres in the last few years with the subsequent increase in patients needing PDA device closure later in life. The anatomy of these ducts is usually challenging for percutaneous closure, being usually Krichenko type C or E. Our first choice in this type of anatomy is the AVP II. We reviewed the results of our practice after 2015.
Methods: We performed a retrospective analysis of the results of paediatric patients who underwent PDA device closure using AVP II after 2015 using our cardiac database (Heart suite).
Results: 24 patient's underwent PDA device closure with AVP II in this period. 20 patients (83%) were ex preterm babies. The duct type was E in 17 cases (70%) and C in 7 cases (30%). All devices were deployed from the venous access. The median age at procedure was 14.88 months (3.5-9.6). The median weight was 8.2 kg (5.3-35.9). The mean PDA size at its narrowest diameter was 3.8 mm (2.5-7) and the mean AVP II size chosen was 9.1 mm (8-12). The device diameter to lumen diameter ratio was 2.39. One device was deliberately oversized (ration device/minimal

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

duct diameter 3) and needed to be snared and retrieved after release since it was causing significant aortic coarctation. It was replaced by a smaller device with no complications in the same procedure. There were no periprocedural complications. Complete closure was achieved in 22 patients (91.6%), 3 patients (12.5%) showed mild flow acceleration in the LPA at last echo without need for intervention. No patients hay an embolization.
Conclusions: The AVP II is a safe and effective device to treat challenging anatomies as types C and E ducts, which are especially common in ex preterm patients. The progressive reduction in indication of PDA closure in the neonatal period will potentially make these cases become more frequent in our daily practice. An asymmetrical version of the device could further reduce the risk of LPA stenosis.

P-174
Long term antihypertensive medication after effective stent implantation in Aortic Coarctation
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Introduction: Aortic coarctation (AoCo) patients frequently maintain hypertension. We assessed determinants of freedom from medication at long-term follow-up after stent implantation.
Methods: We studied 75 patients with native AoCo and recoartation who had undergone effective stent implantation with a follow up of 1 to 17 years. Medication, imaging measurements, Doppler and invasive data were studied.
Results: Native coarctation was present in 47 patients (63%); median age at stent implantation was 25 (SD 15.4) years. Before stenting 66 patients (88%) were on antihypertensive therapy, with 41 (62%) on multiple drugs. Minimal diameter of coarctation was 6.6 (SD 3.7) mm, 25 patients (33%) had a diameter of transverse aorta/aorta at diaphragm level (Tao/DiaphAo) <0.8. Invasive gradients decreased from 42.6 (SD 22.1) mmHg to 5.1 (SD 7.2) mmHg. A second procedure was performed in 12 patients (16%) for multistage procedure (n=6), recoil (n=4), stent fracture, neointima hyperplasia. There were no major complications. At a mean follow up of 7.3 (SD 4.6) years, one patient died of stroke 4 years after the procedure.
Nine patients were not medicated before stenting and remained medication free at follow up. It was possible to discontinue at least one antihypertensive drug in 45 (60%) patients and 21 (28%) became medication free at late follow up. Logistic regression was used to determine predictors of freedom from medication. Patients who became or remained medication free were younger (23.0 versus 32.9 years, p=0.011), had a lower Doppler gradient (38.1 vs 52.8 mmHg, p=0.01), and lower invasive gradient before intervention (32 vs 49 mmHg, p=0.004). In patients with Tao/DiaphAo>0.8, 46% were medication free at last follow up, but with Tao/DiaphAo<0.8 only 20% did not require medication (p=0.015).
A multiple logistic regression model predicted freedom from medication using age, invasive gradient and Tao/DiaphAo>0.8 (AIC 82, p<0.05 in all β-coefficients).
Conclusions: Percutaneous stent implantation in patients with coarctation reduces long term need for antihypertensive medication. Reintervention was required in 16% of the cases. Patients who became and/or remained medication free were younger at the time of stenting, with lower initial gradients and larger Tao/DiaphAo ratio.

P-175
Transcatheter Closure of Aorto-left Ventricular Tunnel in a Symptomatic Infant
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Introduction: Aorto-left ventricular tunnel (ALVT) is a rare malformation. Depending on the size, it results in early and severe left ventricular volume loading and heart failure. Surgery is the treatment of choice, but catheter closure may be considered in limited-resource settings.
Methods and Results: A 2 month old female infant (weight 3.0 kg) presented with symptoms of severe heart failure. Clinical and echocardiographic evaluation confirmed a large ALVT, measuring 9mm at the aortic end, and 6.5mm at the LV end, with a minor constriction (minimum diameter 6.0mm) proximal to the LV opening. The LV and ascending aorta were markedly dilated. In view of the patient's poor clinical condition, transcatheter closure of the ALVT was attempted. Under general anaesthesia, and following a bolus (50units/ kg) of heparin, the defect was crossed retrogradely from the right femoral artery using a 4F MP catheter. An extrastiff guidewire (260 cm long; Cook USA) was advanced into the ventricle, followed by a 6F Amplatzer delivery sheath (St.Jude, USA). Manipulating the guidewire in the LV resulted in ventricular fibrillation, requiring DC cardioversion. Subsequently a 6/6 Amplatzer ADO II device (St.Jude, USA) was delivered to the defect. In the absence of ECG changes suggesting coronary artery compression, the device was released. An immediate improvement in the diastolic blood pressure was noted. The patient was discharged from hospital 48 hours later, and has continued to show clinical improvement at follow-up of 2 months. Follow-up echocardiography shows no residual shunt, and mild, central, aortic valve regurgitation.
Conclusions: In selected cases, in the absence of safe surgical options, catheter closure of symptomatic ALVT should be considered.

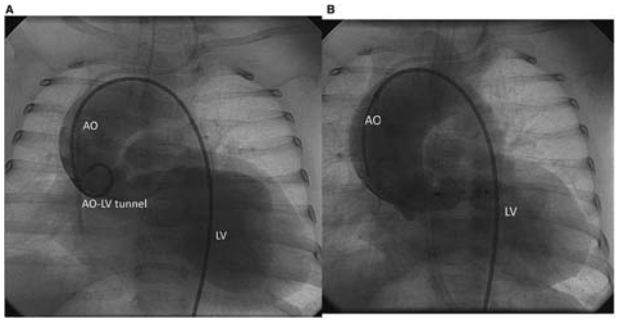

Figure1.
(a) Aortography in anteroposterior projection showing the large aorto-LV tunnel, (b) Aortography after device closure of the tunnel.

P-176
Transcathter closure of patent duct arteriosus (PDA): 62 cases
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Introduction: Trans-catheter closure of patent duct arteriosus (PDA) is new technique that is currently developing in our Paediatric Medical and Surgical Unit.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Objectif: Prove the safety of this intervention that should be a priority rather than a choice, identify the challenges and limitations of this technique.
Material and methods: This retrospective study was carried from October 2013 to July 2017, during which 62 patients who underwent cardiac catheterization in an attempt to close the PDA by trans-catheter approach using duct occluder device.
Results: The average age of our patients was 6.6 years, ranging from 5 months to 15 years. Sex ratio M / F was 0.56. Parental consanguinity is noted in 15 patients. The patients were weighed between 5 kg and 45 kg, and their mean weight was 15.4 kg. PDA presented as an isolated lesion in 84% of patients. PDA-associated heart lesions were as follows: seven cases of restrictive interventricular communication, two cases of pulmonary stenosis. The clinical symptoms in reported cases in our study are very variable, dominated by dyspnea, feeding difficulties, and repeated lower respiratory tract infections.
According to the classification of Krichenko: 37 patients had conical type (type E), six patients had megaphone form (type A), four had window (type B), eleven had tubular (type C), three had aneurysmal (type D). The diameter of the PDA in our cases ranged between 2 mm and 12 mm. Pulmonary hypertension was found in 13 cases. We report three Unsuccessful attempt to close the tubular shape of PDA.
Conclusion: Results of trans-catheter occlusion of PDA have been excellent, and follow-up generally excellent. Trans-catheter closure of PDA deserves a place in the management of this defect and will prove to be of great benefit for all of our patients.

P-177
Few risk factors associated with major adverse events following atrial septal defect closure

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Objectives: Atrial septal defect (ASD) is one of the most common congenital heart disease. Adverse effects following ASD closure are rare. The aim of this case-control study was to study neonatal and pre-interventional risk factors for major adverse events up to one year after ASD closure.
Methods: This retrospective case-control study included all children born in Sweden with ASD closure (surgery or percutaneous device closure) in Stockholm and Lund, between the years of 2000-2014. Cases were children with major adverse events and controls were children without major adverse events occurring during and within one year after ASD closure. Data was retrieved from medical records and the Swedish National Birth Register. Conditional logistic regression was performed to evaluate the association between neonatal and peri-interventional risk factors and major adverse events after ASD closure. Maximum-likelihood estimates of the odds ratio (OR) and 95% confidence interval (CI) was obtained, taking into account potential confounders. The analyses were performed for three categories:

1_ All ASD closures
2_ Children with percutaneous device closures of ASD
3_ Children with surgical ASD closure

Results: Overall 413 children underwent ASD closure at the two paediatric cardiac centers, and 49 (12%) of them were cases with major adverse events during and following the closure. The cases were younger, had less body weight and had a lager ASD size-to-weight ratio compared to children with no major adverse events. There were no neonatal risk factors associated with major adverse events following ASD closure. Pre-interventional cardiopulmonary symptoms was associated with an increased risk of major adverse events for all ASD closures (OR= 2.80 (CI 95% 1.23-6.37), and with an even greater increased risk associated to surgical ASD closure (OR= 4.50 (CI 95% 1.47-13.80). Peri-interventional arrhythmias which needed treatment was the most common major adverse event.
Conclusion: Pre-interventional cardiopulmonary symptoms was associated with an increased risk of major events during and following ASD closure, especially for surgical repair. There were no neonatal risk factors associated major adverse events. These results indicates the necessity of attentive post interventional follow up, maybe even for a longer period of time.

P-178
Novel techniques limit the exposure of children to ionizing radiation related to electrophysiology and ablation procedures

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Introduction: The number of electrophysiology and ablation procedures in children rise in the recent years and so is the related risk of exposure to ionizing radiation. The ALARA rule requires to limit the risk to the necessary minimum. The technological advancements are promising means to the goal. We aim to assess the actual impact of non-fluoroscopic guiding systems (NFGS) during electrophysiological studies (EPS) and catheter ablation procedures (CAP) for the treatment of tachycardia in children.
Methods: We retrospectively analyzed the records of patients undergoing EPS and RFCA. NFGS included EnSiteTM NavX or CARTO® mapping. We analyzed fluoroscopy time, radiation dose area product (DAP). Data is presented as mean ± standard deviation or median (range) dependently on the distribution. Chi-square test, Mann-Whitney and Pearson correlations were employed.
Results: A total of 121 patients and 140 procedures were included with 4 patients a total of three procedures and 11 two. The patients were 15.2 years-old (2.5-18.0) and 76 were male (54%). Forty-four (31%) procedures were done under general anesthesia. The final diagnosis was most commonly AVRT n=69 (55%), AVNRT n=37 (29%) and focal AT n=10 (8%). There were 129 (92%) CAP including 108 (77%) radio-frequency catheter ablations (RFCA), 21 (15%) cryo-ablations (CRYO), and 11 (8%) EPS. In 78 (56%) procedures no NFGS was used, in 46 (33%) NavX, and 16 (11%) CARTO (total of 62, 44%). There was no significant difference in baseline characteristics (age and diagnosis) between no-NFGS and NFGS groups (p=0.407 and p=0.633 respectively) as well as in acute success rate (p=0.404). The fluoroscopy time was significantly longer for CAP 9.0min (0.5–61.0min) vs EPS 3.2min (1.7–6.0min; p=0.022) and so was the radiation dose 5.0 Gy*cm2 (0.02–316.3

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Gy*cm2) vs 1.2 Gy*cm2 (0.1–6.1 Gy*cm2) respectively (p=0.022). The fluoroscopy time was significantly longer for no-NFGS 15.0min (0.5–61.0min) vs NFGS 5.0min (0.7–25.0; p<0.001) and so was the radiation dose 7.8 Gy*cm2 (0.1–118.5 Gy*cm2) vs 1.5 Gy*cm2 (0.02–315.4 Gy*cm2) respectively (p<0.001).
Conclusions: The introduction of NFGS for EPS and RFCA in children significantly reduced fluoroscopy time and radiation dose thus increasing the safety of the procedures.

P-179
Safety of transseptal puncture procedures in pediatric patients weighing ≤15kg
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Objectives: Transseptal puncture (TSP) is a routinely used approach to the left heart in cardiac interventions. The most common complication after TSP is pericardiale effusion (PE). Few data exist on safety of TSP in infants and small children.
Methods: Retrospective data analysis from all patients ≤15 kg who had TSP at our institution from 10/02 to 01/18. We evaluated diagnosis, biometrics, procedure time, fluoroscopic time, and complications. Complications were defined as any incidents attributable to TSP requiring additional diagnostic and /or therapeutical measures beyond standard of care. Procedures with access to the left heart through a patent foramen ovale and patients with other than normal bi-atrial anatomy were excluded. All TSP were performed using standard Brockenbrough needle under fluoroscopic guidance. Biplane X-ray in right anterior oblique (30°) and left anterior oblique (60°) projection was used in all patients.
Results: 23 patients with a body weight of ≤15 kg (female, n=10, 43%) had TSP in a total of 26 cardiac catheterization procedures. 3 individuals had TSP twice. TSP indication was catheter ablation of left sided accessory pathway (AP) in 11/23 (48%) and hemodynamic compromise (such as pulmonary vein stenosis) in 12/23 patients (52%). At TSP, median age was 1.0 (range 0-4.9) years, median body weight was 8.1 (range 1.8-15.0) kg, median procedure time was 215 (range 67-425) min. and median fluoroscopy time was 20.4 (range 3.7-66) min. Patients requiring TSP for hemodynamic reasons were smaller than patients receiving TSP for ablation (6.8 vs. 10.8 kg, p=0.014). 3/12 patients (25%) requiring TSP for hemodynamic reason developed PE after TSP with 2 patients requiring pericardiocentesis/ thoracotomy. In the remaining patient, PE resolved without further therapy. No patient receiving TSP for catheter ablation developed PE. There was no TSP associated death.
Conclusions: TSP in children ≤15 is feasible. However, PE after TSP was associated with lower body weight and the presence of hemodynamic compromise.

P-180
Percutaneous closure of perimembranous and postsurgical ventricular septal defects with Amplatzer Duct Occluder II Additional Sizes in pediatric patients
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Background: Reports of percutaneous closure of perimembranous and residual postsurgical ventricular septal defects (pmVSD and psVSD) are scare. We aim to present our preliminary experience with Amplatzer Duct Occluder II Additional Sizes (ADOIIAS) implantation in selected patients (pts) with pmVSD and psVSD. We have found no previous reports regarding such ADOIIAS application.
Methods: We analyzed retrospectively 6 children with a percutaneous attempt to pmVSD (4 pts) or psVSD (2 pts) closure with ADOII AS.ADO II AS (St. Jude Medical, Inc) is a device originally designed for ductus arteriosus closure.There are three different waist-disc diameters available (3 mm – 4 mm, 4 mm – 5,25 mm, 5 mm – 6.5 mm), every with subsequent three different waist lengths available (2, 4 and 6 mm). Dedicated Amplatzer TorqVue LP 4 French catheter is recommended for the deployment procedure. Briefly, ADO II AS is a symmetrical, self-expanding, single mesh layer nitinol occluder. The devices were implanted in 3 pts from arterial side and in 3 from venous side (after arteriovenous loop creation).
Results: Median age of treated children was 2,5 years (range 1,3 – 8,8). There were 4 aneurysm-type pmVSD (diameter from 2,5 to 3 mm) and 2 psVSD (after Tetralogy of Fallot correction first localized in outflow tract and the second Gerbode type (left ventricle-right atrium connection) with diameter 2,5 and 3,2 mm respectively. There were 4 implants 5/2mm, one 5/4 and 4/4 mm used. All attempts were successful and no major periprocedural complications occurred. The median fluoro time was 25,5 (range 9-36,4) minute. Complete heart block has not been noticed during implantation at any stage of the follow-up (median 10,5 month; range 1-21).We have not observed more than trivial aortic insufficiency after device implantation or tricuspid insufficiency progression. Two patients with psVSD had insignificant residual shunt after the procedure, that was constant in the follow-up.
Conclusion: ADO II AS seems to be a good device for closure selected pmVSD as well as psVSD. Preliminary safety data are encouraging. Mid-term and long-term results should be further evaluated.

P-181
Short and Mid-Term Effects Of Transcatheter Ventricular Septal Defect Closure Treatment On Nutritional Hormones
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Objectives: The aim was to investigate the changes in nutritional hormones of patients before and after the transcatheter closure of Ventricular Septal Defect (VSD).
Method: The study was prospective and case-control and carried out in our Pediatric Cardiology Clinic. 20 VSD patients (13 girls-7 boys, age:2-171 months) and as control group, 26 healthy subjects (16 girls-10 boys, age:3-187 months) were enrolled in the study. Blood samples were collected from all children early in the morning. Insulin growth factor-1 (IGF1), insulin growth factor binding protein-3 (IGFBP-3) and their z scores (sds), insulin, total ptotein, albumin parameters were evaluated. Serum ghrelin and leptin levels were measured using ELISA technique. Laboratory tests and appetite evaluation were repeated at the 1st and 6th month controls in the patient group.
Results: When the initial laboratory parameters of the patients and control group compared; leptin level (median:1981,5 pg/mL) was

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

statistically significant lower than the control group (p=0.010). IGF-1 (median:42.5 ng/mL), IGFBP-3 (median: 2495 ng/mL) and Albumin (median: 4,17 g/dL) levels were significantly lower in the patient group (p=0,027, p=0.018, p =0.035). The ghrelin level (median:1078 pg/mL) was statistically significant higher in the patient group compared to the control group (p=0.037). In the VSD group, IGF-1 (p=0,003), IGF-1 sds (p=0,042), IGFBP3 (p=0,037) and IGFBP3 sds (p=0,030) levels were revealed statistically significant higher at first control according to initial level. Ghrelin and leptin levels increased at the 1st month control then decreased toward 6th month. However, these differences were not statistically significant. Anorexia group (n=9) was compared to non-anorexia group (n=11) with laboratory tests. There was no statistically significant difference. The anorexia was decreased at the first control and was described in only 2 patients. Eight of 20 patients did not come to 6th month control visit. Initial,1st,6th month controls of the 12 patient were evaluated within themselves, a gradual increase of IGF-1 sds and IGFBP-3 sds were determined.
Conclusion: Nutritional deficiency and growth retardation are important problems in patients with VSD. Positive effects on nutritional hormones have been demonstrated in surgical treatments. In our study, we demonstrated the positive effects of transcatheter closure treatment on nutritional hormones.

P-182

Short and Mid-Term Effects Of Transcatheter Atrial Septal Defect Closure Treatment On Nutritional Hormones

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Objectives: The aim was to investigate the changes in nutritional hormones of patients before and after the transcatheter closure of Atrial Septal Defect (ASD).
Method: The study was prospective and case-control and carried out in our Pediatric Cardiology Clinic. 27 ASD patients (14 girls, 13 boys, age:12-197 months) and as control group, 26 healthy subjects (13 girls, 13 boys, age:10-187 months) were enrolled in the study. Blood samples were collected from all children early in the morning. Insulin growth factor-1 (IGF1), insulin growth factor binding protein-3 (IGFBP-3) and their z scores (sds), insulin, total ptotein, albumin parameters were evaluated. Serum ghrelin and leptin levels were measured using ELISA technique. Laboratory tests and appetite evaluation were repeated at the 1st and 6th month controls in the patient group.
Results: When the initial laboratory parameters of the patients and control group compared; ghrelin and leptin level was higher than the control group (p>0,05). IGF-1 sds, IGFBP-3 sds levels were lower in the patient group, but differences were not statistically significant. In the ASD group IGF-1 (p=0,007), IGF-1 sds (p=0,014) levels were revealed statistically significant higher at first control according to initial levels. There was rise in leptin, IGFBP-3, total protein and albumin levels and decrease in ghrelin level but differences were not statistically significant. Anorexia group (n=13) was compared to non-anorexia group (n=14) with laboratory tests. There was no statistically significant difference. The anorexia was decreased at the first control and was described in only 3 patients. Of the 27 patients who underwent transcatheter ASD closure, 7 patients did not come to the 6th month control visit. Initial, 1st, 6th month controls of the 20 patients were compared within themselves; it was found that ghrelin level decreased linearly amoung 6 month follow-up. However, differences were not statistically significant.
Conclusion: Nutritional deficiency and growth retardation are important problems in patients with ASD. Positive effects on nutritional hormones have been demonstrated in surgical treatments. In our study, we demonstrated the positive effects of transcatheter closure treatment on nutritional hormones.

P-183

Interventional treatment in children after TOF correction – late follow up, single centre study

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Tetralogy of Fallot (TOF) is a heterogeneous group of defects with variable degree of right ventricle outflow tract obstruction, hypoplasia of pulmonary arteries and pulmonary blood flow, which influences the strategy for different surgical and interventional procedures. The aim of the study is a retrospective analysis of interventional treatment in children after TOF correction depending on the necessity for initial palliative surgery vs primary correction.
The material consisted of 115 patients (pts) who underwent complete correction of TOF in the period of 2006-2017 in our centre. The data used to analyze were obtained from echocardiography and catheterization (cath) results. All patients were divided into 2 groups: Group 1 (Gr1=36 pts, 31,3%) with initial palliative surgical treatment and Group 2 (Gr2=79pts, 69,7%) with primary complete correction.
The study population consisted of 73 (63,48%) males and 42 (36,52%) females. The median of observation time after correction was 5,34 years with interquartile range (IQR) [2,9 – 8,3]. The median of correction age was 16,3 [4,9-48,5] months, however children in Gr1 had correction significantly later than Gr2 (18,3 vs 12,8 months, p=0,008) with similar early mortality (8,3% vs 7,6%). The medians of z-scores at time of correction were significantly lower in Gr1 for the left (-2,05 vs -0,12, p=0,004) and right (-2,85 vs -1,02, p<0,001) pulmonary artery. In the post-operative period generally 36 percutaneuous interventions single or combined were performed and included: balloon pulmonary valvuloplasty (16 in Gr1 vs 4 in Gr2), pulmonary artery stents implantations (15 vs 5), ASD closure (1 pts). Children in Gr1 after correction significantly more often needed: 1) single interventional cath (50,0% vs 7,59%, p<0,001), 2) at least one implantation of the stent into pulmonary artery (27,78% vs 6,33%, p=0,003), 3) reinterventions (more than 1 cath) (27,78% vs 2,53%, p<0,001) and reoperations (n=11; 30,56% vs n=10;12,66%, p=0,041). Analysis of Kaplan-Meier estimator revealed that probability of reoperation (log-rank p=0,025) and at least one interventional catheterization (p<0,001) is higher in Gr1.
The patients with TOF requiring initial palliative treatment had greater risk of interventional catheterizations after surgical correction as well as reoperations mostly due to initial hypoplastic pulmonary arteries.

P-184

Transcatheter closure of congenital and acquired Gerbode defects with Nit-Occlud Le VSD (PFM) coil. Immediate and mid-term results

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Background: The study describes our experience with interventional treatment of congenital and acquired Gerbode defects using Nit-occlud Le VSD coils. The patients were selected based on 2DE exam. The assessment included a detailed location and dimension of the defect, Qp/Qs ratio, and the relationships with nearby valvular apparatus.
Material/methods: The procedure was performed between 31.10.2014 and 14.01.2016 in seven children (4 F, 3 M), including one infant with native, and six older children with acquired, post-operative LV-RA shunts: in one after correction of ASD1 and significant anterior mitral valve cleft, and in six after closure of pmVSD (coexisting with other congenital heart defects, such as: CoAo - in two, IAA type B – in other two, and DORV- in one patient). The age of the patients was: 8 month - 17.8 years (x - 10.2 ±5.4 years), body weight: 7.4 kg - 56 kg (x-35.5±16.4 kg). The diameter of the defects based on the 2-DE examination was verified and comparable to found in the angio-cardiographic study (x - 4.2±1 mm vs 4.1±0.36 mm), Qp/Qs ratio was: x - 1.7±0.3. The procedure time was 40-65 min (x-53.6±10.7 min). The size of the coils used was: from 8x6 to 12x6 mm.
Results: In an infant with direct Gerbode defect shortly after the procedure hemolysis was observed, lasting for three days. This required blood transfusion, therapy with steroids and propranolol. Another patient temporarily experienced increased ventricular ectopy. Immediately after the procedure residual shunt was observed in five patients: mild in one (with direct LV-RA shunt), trivial in four. In two children the defect was completely closed. The follow-up period was 2-36 months (x – 20.7±10 months). Control 2DE study revealed no residual shunt in 6 patients. In one, with longest observation residual shunt was still present (2.7 mm in diameter, below the implant). In another patient serious tricuspid regurgitation co-existing with LV-RA defect, after the procedure decreased to insignificant in the 2-month of follow-up.
Conclusions: Nit occlude le-VSD coils are useful and safe in transcatheter treatment of direct and indirect LV-RA shunts, however require a lot of experience and skill, especially in the youngest patients.

P-185
Five years outcome after neonatal ballon aortic valvuloplasty in critical aortic stenosis including a cohort of prenatally dilated valves

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Background: New surgical valve repairing techniques even in newborns challenge the interventional approach. Prenatally dilated valves add on a new subgroup of patients. We reviewed our results of neonatal balloon aortic valvuloplasty (BAV), whether this method can still be offered as first line treatment.
Methods: Retrospective analysis of all consecutive patients from 2005-17 who received BAV in the neonatal period due to critical aortic valve stenosis as leading lesion. Patients with suspected single-ventricle physiology or without consistent F/U were excluded.
Patients: 34 newborns, 11 of them after intrauterine treatment, median body weight 3.1kg (2.0-4.3), median valve diameter 6.6mm (overall z-score -0.3, in the non-fetal group +0.18, in the fetal group -1.1). Accompanying lesions were EFE in all of the fetal and 3 in the non fetal group.
Procedure: Median echo gradient was reduced from 78mmHg (20-144) to 43mmHg (17-72). Aortic regurgitation was found trivial or mild in 29, moderate in 4 and severe in 1 patient.
Complications included 2 femoral artery obstructions (treated and completely relieved); 3 femoral vein obstructions, 2 intimal flaps in aortic arch.
No peri-procedural death, no cerebral bleeding nor infarction and no mitral damage could be observed.
Results: Neonatal Ross-Operation was necessary in 6 in the non-fetal and 6 in the fetal cohort. 30 patients met the F/U completion. Five years after BAV 12 patients still are without further treatment (10/22 in the non-fetal group and 2/8 in the fetal cohort), with mild to moderate valve stenosis and/or regurgitation.
Conclusion: Neonatal BAV in critical aortic valve stenosis can be considered as a safe procedure with reasonable risk and complication rate. BAV may avoid early cardio-pulmonary bypass surgery in the neonatal period in the majority and preserve the valve for the first years. Intrauterine BAV seems to create a different subgroup of patients with a more severe disease spectrum where usual strategies for postnatal BAV may not apply and early valve replacement must be considered, especially in small valves and the presence of EFE.

P-186
Tapering of guiding catheters with coronary angioplasty balloons: a new tip to reach hard places

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Introduction: Abnormal vascular communications are commonly observed in patients with congenital heart disease and may contribute to deterioration of patient clinical status. Treatment can be performed either by surgery or by percutaneous techniques. Transcatheter closure has some advantages over surgery, including shorter recovery time and avoidance of thoracotomy, however it may not be feasible due to anatomical or technical issues.
Methods: Two patients underwent percutaneous closure of vascular malformations. A 17-year-old male with medical history of pulmonary stenosis and complaints of progressive dyspnoea and fatigue, was diagnosed as having a large coronary artery fistula (CAF) arising from the left main coronary artery to the left atrium. A 46-year-old female with previous surgery for scimitar syndrome and recurrent haemoptysis due to a major bronchial artery arising from the descendent aorta to the lower right lung lobe. Both abnormal vascular connections had a tortuous arterial course, with ostial narrowing and extensive angulation.
Results: In both cases, multiple attempts to selectively catheterize the afferent vessel with an appropriately sized 5Fr delivery catheter were unsuccessful. To overcome the narrow ostium and significant angulation, a coronary angioplasty balloon was used to taper the guiding catheter distal extremity thus creating a more favourable shape to overcome the lesions stenotic entrance and tortuous course. In both cases, this manoeuvre allowed for an easy advancement and positioning of the guiding catheter. The CAF was successfully occluded with an Amplatzer Duct Occluder II Additional Size and the bronchial artery was also successfully closed with an Amplatzer Vascular Plug II. There were no complications during both procedures.
Conclusions: There has been several advances in transcatheter procedures over the last decade with development of various innovative tools. These cases describe a new percutaneous technique to overcome the difficulties of gaining access to complex vascular malformations, through a stenotic angulated entrance, which would otherwise have to be treated surgically.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Early Experience with the Bentley Be-Graft Stent in Treatment of Aortic Coarctation and Pulmonary Artery Stenosis in Children

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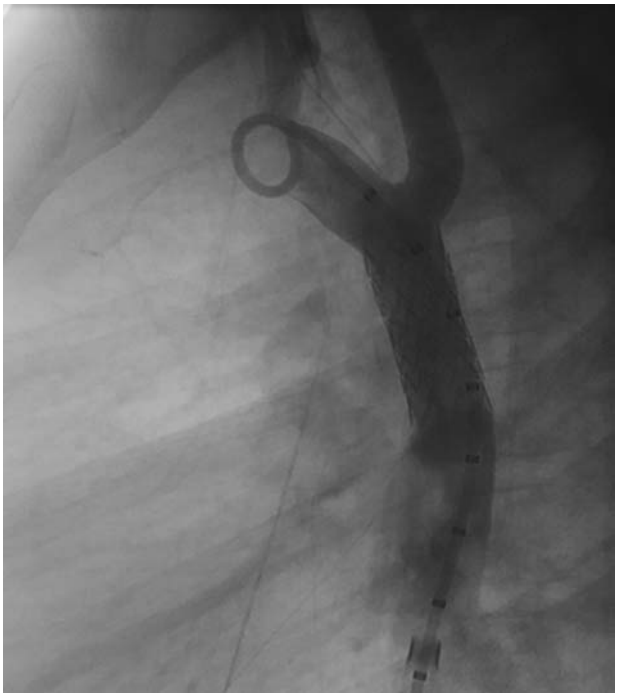
Introduction: Transcatheter stent therapy for aortic coarctation in older children and adolescents has over the past decade become a frequent treatment strategy of choice. Favorable properties of stents for coarctation include 1) high radial strength at maximal inflation diameter, 2) limited foreshortening, and 3) low profile. The Bentley BeGraft aortic stent arrived on the European market in 2016 and is a premounted, balloon expandable, covered stent indicated for the use of native or recurrent CoA. Here we report our first eight pediatric cases of using this stent for aortic coarctation and one case of pulmonary artery stenosis.
Methods: Between November 2016 and November 2018 Bentley BeGraft stents were implanted at our center in aortic coarctation of eight patients with native membranous aortic coarctation near the isthmus. Mean age was 9.9 years (range 4.0-14.3) and mean weight 38.0 kg (range 19.4-58.7). All patients had pre-procedure CT or MR imaging. Acute outcome evaluation was based on angiography (Image) and change in coarctation pressure gradient. Short term outcome was evaluated by clinical assessment and follow-up CT scan and chest x-ray performed 4-6 months after stent implantation. Placement of the stent in the pulmonary artery was performed in a 4 year-old TCPC-operated single ventricle patient.
Results: Invasively measured coarctation gradients ranged 16-31 mmHg (mean 25) and were reduced to 3 mmHg or less in six of eight patients. Stents that were inflated to nominal pressure reached their expected diameter (12 or 14 mm). There were no acute or short term complications related to the procedure. There was minimal to no recoil and no fracture at 4-6 months follow-up. All patients were clinically well at follow-up. Follow up results on the pulmonary artery stent is expected shortly.



Conclusions: Based on our initial clinical experience this has a high ease of use with a rapid operator learning curve, favorable low profile allowing for uncomplicated use in patients 20 kg and above, and it shows minimal recoil at 6 months. In summary, our early results support that the Bentley BeGraft stent is suitable for use in aortic coarctation in older children and adolescents.

P-190

Percutaneous Transhepatic Valvuloplasty of a Trimmed Melody Valved Stent in Mitral Position in a 2-Year-Old Infant

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Introduction: This report describes the first case in our knowlegde of a patient with a Melody valve implanted in mitral position in which a posterior valvuloplasty was performed by transhepatic and transeptal approach.
Case Report: Male infant antenatally diagnosed with critical aortic stenosis. After three different surgeries including a Ross operation, a mitral valve repair and a mitral valve replacement with a mechanical prosthesis, he underwent a Melody valve implantation in mitral position at the age of 6 months.
During follow up, the patient developed severe stenosis of the Melody valve with a mean gradient of 10 mmHg. Therefore a percutaneous balloon valvuloplasty of the prosthesis was performed at the age of 2 years and 9 months (weight 10.5 kg). Due to a previous complete thrombosis of both venous femoral systems and the presence of an intact interatrial septum a transhepatic procedure with transeptal puncture was chosen as a feasible approach for this patient.
The valvuloplasty on the Melody prosthesis was performed using a 18 and 20 mm balloons, resulting in a significant decrease in the mean gradient of the valve. Hemostasis of the transhepatic access was achieved by embolizing the tract with a 6 mm Amplatzer Vascular Plug IV (SJM) and manual external compression of the skin at the puncture site. There were no complications and the patient could be discharged the next day. On the predischarge echocardiogram, mean gradient across the valve had decreased down to 6 mmHg without an increase in the degree of the valve insufficiency.
Discussion: In our patient, in order to avoid a fourth extracorporeal surgery, percutaneous approach was proposed as the technique of choice.
Due to the previous history of this patient, venous femoral accesses were already thrombosed and unable to be used for the procedure. In this situation, transhepatic access is a very useful and feasible alternative.
In our case, the procedure was performed by transhepatic approach and transseptal puncture with no technical complications and good hemodynamic results.

P-191

Percutaneous coronary intervention in children: a single center experience

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Objectives: To investigate the feasibility, procedural techniques, safety, and overall potential of percutaneous coronary angioplasty

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

and stent implantation in infants with coronary artery disease and acute coronary syndrome.
Methods: Retrospective review including all patients under 18 years old who underwent percutaneous coronary intervention during a period of 14 years.
Results: Between 2004 and 2018, 15 patients underwent coronary intervention, including percutaneous coronary balloon angioplasty in 2 and coronary stent implantation in 13. Median age was 5,5 years (range 13 days–17 years), and median weight was 17 kg (range 3,3–90). Indications for intervention included postoperative acute coronary syndrome in seven cases (46%), in the setting of arterial switch (3), Ross (3), and anomalous coronary (1) surgical procedures; severe coronary allograft vasculopathy in heart transplant recipients in four cases (26%); late severe ostial stenosis post arterial switch operation in two cases (13%); and acute coronary syndrome associated with Kawasaki or Wlilliams syndromes in two cases (13%). Successful stent placement with excellent revascularization was achieved in all cases, with an average internal diameter of 2.7 mm (ranging from 2,25 to 3,5 mm). Balloon angioplasty alone resulted in optimal resolution of coronary stenosis in 2 patients under 2 months of age. Seven patients were in ECMO support during the procedure; four of them died in ICU postprocedure period. There were no late deaths (overall survival of 74%). Average intervention-free period in survivors was 3,5 years; severe in-stent re-stenosis ocurred in 1 case and was succesfully percutaneously treated at 4,1 years of follow-up.
Conclusions: In our experience percutaneous coronary intervention and stent implantation is a feasible and safe option in infants with coronary stenosis. Short-term benefits of the technique can be critical in certain cases, and it is a viable strategy for bridging patients with acute ischemia or poor ventricular function to elective surgical revascularization or transplantation.

P-192
Transcatheter closure of atrial septal defects in paediatric patients with the Nit-Occlud ASD-R device
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Objective: To describe our experience with the Nit-Occlud ASD-R (NOASD-R) device for percutaneous closure of ostium secundum atrial septal defects (OS ASD) in a paediatric population.
Methods: Retrospective observational study.
Results: From Feb 2014 to Nov 2018 sixty-six patients underwent attempted transcatheter OS ASD closure with the NOASD-R device. Implantation was successful in 65 patients (98.5%). In one of the patients a 20 mm device was deployed across the defect but the left disc did not expand adequately so it was not delivered. 41 patients were female with a median age of 5 years [ICR:3.7-9] and a median weight of 19.7 Kg [ICR:15.5-28.5]. The mean ASD diameter by TOE was 11.5±3.7 mm. Eight (12.1%) patients had multiple defects and the aortic rim was deficient in 19 (28.8%). The mean 2D diameter/weight ratio was 0.59±0.3 (median 0.53, ICR:0.35-0.82). The median fluoroscopic time was 7 min [ICR: 5-11.7]. The median size of the devices was 14 mm [ICR:10-16]. The mean device size/2D diameter ratio was 1,19±0.18. Additional intervention was required in 4 (6.1%) patients. There were no major complications during the procedure. Minor complications happened in 4 (6%) patients; one had a supraventricular tachycardia, which resolved after mechanical stimulation of the atrium, 2 patients developed first and second-degree heart block which resolved spontaneously within a week and another patient had a femoral arteriovenous fistula. Twenty (30.3%) patients had low velocity residual flow through the device at the 24 h TTE. Two patients experienced small pericardial effusion after the procedure with complete resolution within a month. At a mean follow-up interval of 27.3±15.7 months complete occlusion was achieved in 64 (97%) patients and there have been no episodes of late embolization, erosion, endocarditis, neurologic events or death.
Conclusions: Closure of OS ASD with the NOASD-R device is safe and effective in the paediatric population. Most patients had short-term intra-device residual flow but the definitive closure rate is high. Implantation success is high with no major complications in the short and medium-term.

P-193
Spanish Registry of percutaneous VSD closure with NitOcclud® Le VSD-Coil device: lessons learned after the first hundred implants
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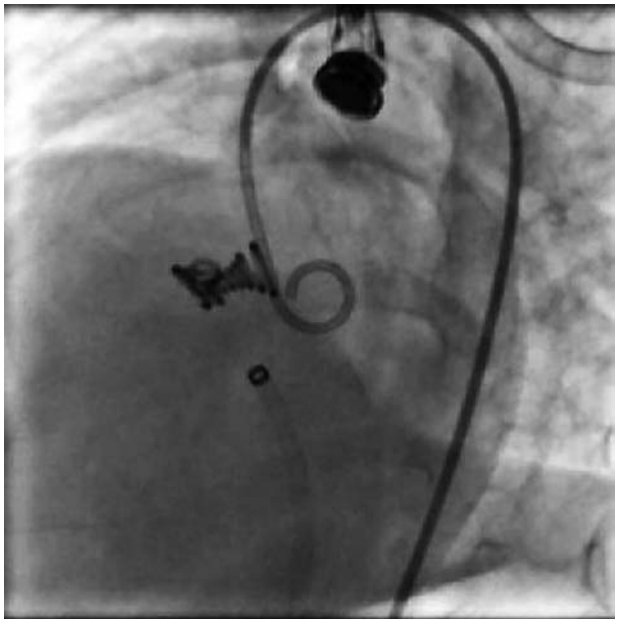
Introduction: The NitOcclud Le VSD Coil was specifically designed for transcatheter occlusion of VSDs and became available for this purpose in August 2010. Our objective is to compile the Spanish experience and present the midterm results of this technique.



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Methods: Spanish multicentric observational study, which retrospectively recruited all patients (any age) with VSD (any location or nature) who underwent percutaneous NitOcclud occlusion of their defect, using an intention-to-treat analysis, until May 2018.
Results: 105 attempts were made to implant at least NitOcclud® in a total of 104 patients, whose procedures were performed in 9 institutions (representing more than 96% of the national experience). The median (range) of age and weight was 8.6 years (0.4-68) and 25 kg (5.8-97) respectively. Ten patients were <9Kg. The VSD was an isolated defect in 88 of the cases. The classification by its septal location was: 86 perimembranous (69 with aneurysm), 5 muscular, 4 Gerbode and 10 related to a surgical patch.The mean fluoroscopy time was 37 '. The implant was successful in 96 of the 105 procedures and its (range) follow-up time was 1.8 years (0-4.5). Of these, 4 had to be explanted due to severe haemolysis (n = 2), embolization (n = 1) or transient complete AV block+ significant residual shunt (n = 1). In the others (n = 92), the procedure was safe, without major complications, and the initial percentage of complete occlusion of the defect without residual shunt or in a minimum degree was 71% (68/96) (complete occlusion= 32, trivial shunt= 36) and final percentage of 90% (83/92) (complete occlusion= 62, trivial shunt= 21). Four patients required a second procedure for residual shunt occlusion with additional devices. There were no deaths and the percentage of total hemolysis was 4.7% (2 of them with spontaneous resolution).
Conclusions: The NitOcclud® device can be used successfully for a wide variety of selected patients with VSD. We have not found permanent changes in AV conduction. Patients with residual shunt should be periodically checked to rule out the occurrence of clinically significant haemolysis and delaying the start on aspirin after discharge is our recommendation.

P-194
Clinical features, management and outcome of Infective Endocarditis after Transcatheter Pulmonary Valve Implantation

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Introduction: Transcatheter pulmonary valve implantation (TPVI) is a valuable treatment option for patients with dysfunctional right ventricle outflow tract (RVOT).
Infective Endocarditis (IE) has been reported as an important complication of this procedure with potential lethal consequences. We aimed to review the clinical, features, management and outcomes of IE after TPVI in a tertiary hospital centre.
Methods and Results: Retrospective review of the clinical records, and imaging, of the patients diagnosed with IE after TPVI (Melody/Edwards Sapien) in our Congenital Heart disease Unit between 2012 and 2018.
Results: From February 2012 to November 2018, 46 patients (24 males- 52%) underwent TPVI (33 Melody, 13 Edwards). 6 patients (13%) had been diagnosed with 7 IE episodes. One of the patients, with Down Syndrome and chronic dermatitis had a second episode of IE 2,5 years after the first episode was resolved. IE diagnosis was based on clinical symptoms, positive blood cultures in 6 episodes, with one episode with 2 different species found: Staphylococcus Aureus MS (N=3), Streptococcus Sanguis (N=1), Streptococcus Anginosus (N=1), Streptococcus Coagulasa Negative (N=1) and Streptococcus Parasanginis (N=1). Positive signaling of the TPVI in the PET/CT was found in 2/7 when echocardiogram was not conclusive. Echocardiography showed vegetations in 2/7 IE episodes, but in all an increase in the Doppler gradient across the valve and stent. All patients received intravenous antibiotic treatment according to the antibiogram for at least 6 weeks except one patient who received 8weeks antibiotics therapy. None required surgical explantation or percutaneal reintervention on the valve. 5 patients were discharged with a mildly dysfunctional valve (mild regurgitation and stenosis), and with normal valve function in 2 patients (no regurgitation and peak gradient <30mmHg). As predisposing conditions for IE, dental procedure weeks after IE (n=1), Chronic dermatitis (n=2) and 2 previous IE in the conduit before the TPVI (n=1). All had been on regular aspirine treatment after the TPVI. There were no deaths.
Conclusions: In our serie, all IE were cured with antibiotic treatment, leaving mildly dysfunctional valves. None of the patients required surgical/percutaneous interventions, either in the acute episode nor in a mid term follow up.

P-195
Tailored approach to transcatheter palliation of critically reduced pulmonary blood supply. Data on long term follow up

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Introduction: Proper development of pulmonary tree is the single most important determinant of the outcome of patients with congenital heart disease and critical pulmonary blood supply. The most appropriate algorithm of treatment is still a matter of debate, in particular regarding the role of trans-catheter palliation. We present the long term outcome of tailored transcatheter palliation of this group of patients.
Methods: We retrospectively analysed data on patients who had undergone trans-catheter palliation of duct dependant congenital heart disease Between 2005 and 2017 in our institution. Arterial duct or RVOT PTA /stenting were performed according to a prespecified algorithm.
Results: Overall 47 patients underwent either AD stenting (N=42) or RVOT PTA/Stenting (N=5). Median age at catheterization was 13 days (3-686). Median weight was 3 Kg (1,5-4). There was one procedural death (2%). The procedure was unsuccessful in 8 patients (10%). Mean follow up was 1127±886 days. Eleven patients out of 46 died (23,9%). One year, five year and ten year survival were 82% (95% CI 68-90); 75%(95% CI 60-85); 60% (95% CI 27-81), respectively. At Cox regression analysis weight and univentricular physiology remained independently associated with long term mortality, HR 0.3 (95% CI 0.12-0.88), p=0,03; HR 5 (95% 1,3-20,2) p=0,02, respectively. In the whole population 23 patients (50%) needed further intervention before surgical repair or definite palliation, overall median time to reintervention was 146 days (83-1087). Central pulmonary arteries grew in all patients but to a much higher extent in patients submitted to RVOT stenting.
Conclusions: Tailored trans-catheter management of congenital heart disease associated with critically reduced pulmonary flow resulted in proper maturation of pulmonary tree allowing surgical repair or definite palliation. Patient with severely hypo plastic pulmonary arteries should be elected to undergo antegrade pulsatile flow restoration.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-196

Diagnosis and outcomes of patients with supravalvular pulmonary stenosis treated with percutaneous balloon dilation

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Introduction: Congenital supravalvular pulmonary stenosis (PS) is a rare subtype of PS. Outcomes following percutaneous dilation of the pulmonary artery (PA) and pulmonary valve (PV) seem to be dependent on the presence of a supravalvular component. The aim of this study was to determine the reliability of echocardiography in identifying the origin of the PS and to evaluate the efficacy of percutaneous dilation of supravalvular stenosis.
Methods: Retrospective study of 109 patients having undergone percutaneous dilation of a native PS between 2006 and 2017 in a tertiary pediatric hospital. Patients were classified as having supravalvular PS or non-supravalvular PS based on angiography. Echocardiographic measurements (main PA and PV diameters) and interventional results (hemodynamic and echocardiographic right ventricular (RV) to PA gradients) were compared between groups.
Results: Mean age at intervention was 1.93±4.22 years old. Angiography identified a supravalvular component in 26 patients (24%). Anatomical description of the PV and PA by echocardiography had a sensitivity and a specificity of 58% and 84% respectively. Independently of the coexistence of another type of PS, patients with a supravalvular component had a significantly smaller PA:PV ratio measured by echocardiography: 0.84±0.30 vs 1.47 ±0.54 ($p<0.001$). Based on the PA:PV observation, ROC analysis determined an area under the curve of 0.851 (95%CI 0.739-0.963) and a discriminative cut-off ratio of 1.048 according to Youden's index; increasing both the sensitivity and specificity to 86%. After dilation, hemodynamic gradients were reduced from 37.35±20.51 mmHg to 26.04±12.84mmHg in patients with a supravalvular component compared to a decrease from 39.80±20.47mmHg to 11.75±8.67mmHg in patients without ($p<0.001$). The gradient remained significantly higher among patients with a supravalvular PS at the 6-12 month follow-up: 48.94±28.75mmHg compared to 22.54±21.28mmHg ($p<0.001$). Of patients with supravalvular stenosis, 19% required a second intervention (percutaneous or surgical) compared to 6% of those without a supravalvular component ($p=0.04$).
Conclusions: Systematic calculation fo the PA:PV ratio significantly improves the accurate determination of the origin of the pulmonary stenosis compared to anatomical description alone. Percutaneous dilation of a supravalvular PS is less effective and patients are more likely to require a second intervention than patients with pulmonary stenosis without a supravalvular component.

P-197

Clinical implementation of a model-based training course to reduce the learning curve in interventional therapy of congenital heart defects

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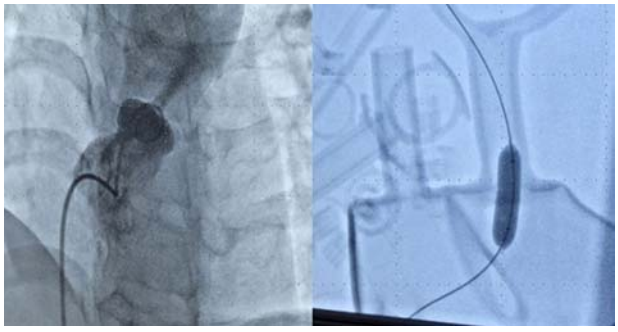
Introduction: Cardiac catheterisation has become a routine alternative to open heart surgery for many aspects of congenital heart



defects. Today, many "simple" defects such as PDA, ASD, pulmonary or aortic stenosis or coarctation of the aorta are managed interventionally whenever suitable. Despite the state-of-the-art technical equipment in most of the cardiac catheter labs, there has not been much change in the practical training. Training is still performed under the expert guidance of an experienced doctor mainly by "practicing on the patient". As the number of cardiac interventions is steadily increasing and the interventions become more complex, the interest for new training opportunities and modalities increase.
Methods: After developping variuos 3D silicon heart models we established a comprehensive clinical training course for PDA closure and balloon dilatation of the pulmonary valve and implemented this model based course as a part of two one week workshops at the Ayder Hospital in Mekelle, Ethiopia. The pediatric and cardiologists participating in the workshop have not carried out interventional catheterisation before. The course consisted of an initial full day model based training followed by actual treatment of patients with PDA and pulmonary stenosis. At the beginning of every day a repeat model catheterization was performed. In order to evaluate the efficacy of these models, the required X-ray time for the interventions was measured at different times.
Results: After repeat hands-on training 17 patients (15 PDA, 2 PS) were treated successfully by the newly trained cardiologists as first operators. Handling of the catheter material including wires, balloons and catheters was smoothly. All new investigators confirmed the realistic aspect of the models. There was a significant reduction in the fluoroscopy time from the beginning (40 minutes) to the end of the workshop (18 minutes).
Conclusions: Real size 3D models could offer an attractive alternative in the education and training of interventional cardiologists. Real beginners could get the possibility to practice unlimited time on the model and improve their practical skills in order to be possibly best prepared for treating real patients. In addition these models may be suitable for maintaining technical skills in lower volume centers.

P-198

Establishment of a 3-D silicone model to facilitate realistic hands-on-training for diagnostic and interventional pediatric cardiac catheterization

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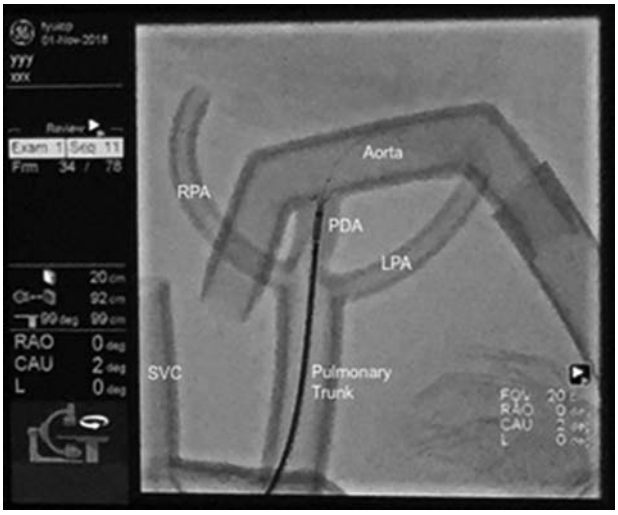
Introduction: Today, cardiac catheterization is still of great importance for diagnostic and therapeutic purposes in pediatric cardiology. Nevertheless there are currently no widely accepted training

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



models for trainees or junior doctors to learn these techniques. Especially the manual management of long catheters and wires as well as balloon catheters and exchange wires may remains a hurdle in the rapid development of catheter skills.
Methods: We developed various silicon heart models including a normal heart and models with common congenital heart defects such as a patent ductus arteriosus (PDA), pulmonary and aortic stenosis, coarctation of the aorta and ASD. The models were built out of silicon tubes, then CT-scanned and finally printed with a 3D-Silicon printer.
To test the effectiveness of the models, a comparative training course with a manual was implemented. This training course was then offered to medical students with no previous catheter experience, residents and fellows with no and/or little catheter experience and pediatric cardiologists with high experience including interventional catheter treatment. After initial theoretical teaching, practical hands-on training was performed. There was a stepwise escalation in the skills required from purely diagnostic catheter investigation including hemodynamic assessment up to interventional therapy such as balloon dilatation of an aortic or pulmonic stenosis or coarctation stenting. The improvement of skills was assessed by timing of each procedure.
Results: After a short training period the training models enabled even completely unexperienced trainees to handle the fluoroscopy machine and catheters, wires, balloons and stents. The acceptance of the model by experienced cardiologists was high.
Conclusion: These preliminary results show that – like in other fields of medicine or industry – training models may help to improve technical skills and reduce a learning curve in managing complex procedures. Implementing a model based training of catheterization procedures might have a lasting learning effect on cardiologists. A reduction of hands-on training in animal models may be possible; even for experienced staff, a model based training in the handling of devices may be helpful before intervening in patients. We believe that model training should be further implemented in the future training of cardiology residents.

P-199
The risk of infective endocarditis following interventional pulmonary valve implantation. A Meta-Analysis
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Background: Interventional percutaneous pulmonary valve implantation (PPVI) was first reported in 2000. Today two different valves are certified for this procedure (i.e. Medtronic Melody® valve and Edwards Sapien™ valves): the procedure became commonly available and was increasingly used from 2010 onwards. For a decade studies have reported an increasing risk of infective endocarditis (IE) after PPVI; patients for PPVI are usually younger and with decades of lifespan to come, therefore even a low annual incidence of IE is important. The overall incidence and a potential difference between the valves however remains unclear.
Methods: A systematic literature search was performed in the databases Medline, Cochrane Library and Embase including the clinical trials register. The time period was between 01/2000 and 12/2018. The aim was to summarize and compare the cumulative incidence of IE after PPVI. In addition, at a sensitive analysis we set the incdence rates of the two valve types in ratio with a normal population.
Results: A total of 967 publications were identified searching for „pulmonary valve implantation", „PPVI", and 47 publications were used for final analysis. 3616 patients with Melody® valves and 501 with Sapien™ valves were included. IE after PPVI occurred in 214 patients with Melody® valves and in 5 patients only with Sapien™ valves. The pooled incidence for Melody® and Sapien™ valves was 4.9 percent (95% CI: 3.6 – 6.2) and 1.3 (95% CI: 0.3 – 2.3) respectively. Chi-square test was significant. The sensitivity analysis showed that the incidence rate ratio was 252.1 (95% CI: 187.6 – 338.6) for Melody® valves and for Sapien™ valves 2.7 (95% CI: 0.8-9.2).
Conclusions: PPVI is the treatment of choice wherever feasible; since all biological valves have a limited lifespan and due to the relatively young age of the majority of patients, the cumulative risk of IE is of particular importance. Based on the data presented, one of the two catheter valves currently available clearly is superior with regard to the risk of BE. The reason for this is unclear, but it seems likely that the different biological material as well as the mode of preparation may influence this outcome.

P-200
Interventional cardiac catheterization in neonatal age: results and early outcome in a multi-center Italian experience
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Introduction: Interventional cardiac catheterization in newborns is a challenging therapeutic option. However, technical advances and improvement of cardiologist's expertise widened indications and complexity of this approach in these patients.
Methods: Interventional cardiac catheterization procedures performed in neonatal age in all Italian high-volume referral centres

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

of Paediatric Cardiology were analysed in terms of morbidity and procedural or in-hospital mortality.
Results: From January 2000 to December 2017, 1238 newborns underwent 1353 percutaneous cardiac interventional procedures. Mean weight was 3.00±0.55 kg (range 1.00-5.80) and median age was 2.0 days (range 0-31 days). Mortality and morbidity of this approach were 6.7% and 9.5%, respectively. At multi-variate analysis, in-hospital mortality was significantly related to low-weight at procedure ($p<0.01$), procedural failure ($p<0.01$), univentricular physiology ($p<0.01$) and genetic syndromes ($p<0.01$). A trend to increase of number of the procedures was recorded over time (38 procedures/year in 2000 vs 90 procedures/year in 2017). In addition, also an increase in complexity of the interventions was reported over time. In fact, Rashkind atrioseptectomy and balloon valvuloplasty (either aortic or pulmonary balloon valvuloplasty) decreased from 81.6% of all procedures in 2000 to 71.1% in 2017 ($p=0.3$). This trend was presumably due to an improvement of techniques and materials which allowed different and more complex interventional procedures. These changes increased procedural complication rate and in-hospital mortality over time. These increases were more evident from 2000 to 2013 (both complication and mortality rate increased from 2.6% to 12.3%, $p=0.08$). However, from 2013 to 2017, no significant change in number and type of the procedures was recorded, although with significant decrease of failure (from 7.5% to 2.2%, $p=0.09$), complication (from 12.3% to 4.4%, $p=0.05$) and mortality rates (from 12.3% to 3.3%, $p=0.02$), respectively.
Conclusion: Interventional cardiac catheterization is a safe and feasible alternative to surgery in neonatal age. Low-weight, univentricular physiology, associated syndromes and the failure of procedure significantly contribute to procedural and/or in-hospital mortality. However, future improvements in techniques and operator skills as well as a more accurate selection of patients might improve results and early outcome of this approach in this high-risk subset of patients.

P-201
Results of catheter interventions on ECMO circuit

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Objectives: To describe our institutional experience of performing cardiac catheterisation including intervention whilst on ECMO circuit.
Methods: All patients <16 years who underwent cardiac catheterisation whilst on ECMO between November 2010 and November 2018 were identified from the departmental database and a retrospective case note review carried out.
Results: During the study period 41 patients had 47 angiography/intervention on ECMO, of whom 11 had 14 procedures carried out via the arterial limb of the circuit the remaining had catheterisation using normal access.
The ECMO circuit is accessed by using a Y connector cut into the arterial ECMO tubing with an incorporated haemostatic valve through which the catheters are passed for angiography and interventional procedures.
Patients ages ranged from 1 day to 14 years (Median 9 months) with average weight of 15.7 kg (range 2.4-71.8).
Of the 41, 17 were univentricular (1 no palliation, 7 BT shunts, 6 Norwood stage 1,2 PA bands, 1 Nikkiadoh) and 7 had had biventricular repair (2 Ross procedure, 2 arterial switches, 1 double switch, 1 Fallot repair). Eight had cardiomyopathy, 8 had a heart transplant (4 cardiomyopathy, 1 TGA, 1 Shone complex and 1 HLHS). One had pulmonary hypertension.
41 patients had angiography all of which were diagnostic with 31 having a procedure 3 of which via the ECMO circuit (6 Atrial septostomy, 5 Left atrium decompressions, 4 cardiac biopsies, 3 Pulmonary artery stents, 5 shunt revascularisation stents, 2 Embolisations, and 1 SVC stent). Only one procedure, an atrial septostomy was unsuccessful.
There were no circuit related infections and no complications from procedures. Eight patients could not be weaned from ECMO, the remainder survived to PICU discharge.
Conclusions: Intervention is possible through the ECMO circuit whilst the patient is on ECMO. Access is an important consideration when performing catheterisation on ECMO and can be safely performed through the existing arterial limb of the ECMO circuit, and this provides the necessary access for diagnosis and therapeutic intervention

P-202
Pulmonary arteriovenous malformation embolization with occluder device

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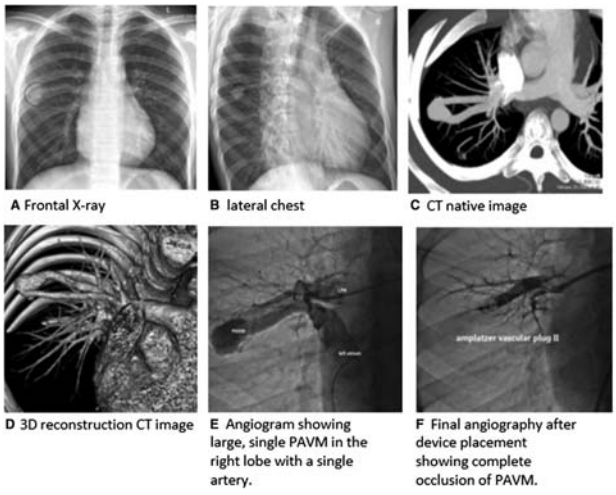
Objectives: to present the case of embolization of the pulmonary arteriovenous malformation (PAVM) with occluder device.
Methods: A 13-years-old boy was admitted with tachypnea, mild cyanosis during physical activity. He underwent chest X-ray that shown nonhomogeneous shadow in the right lung (fig. A, B). CT scan revealed a large PAVM (35x28 mm) in the middle lobe of the right lung (fig. C, D) and patient was admitted to the cath lab (fig. E). Amplatzer vascular plug II 10 mm was placed 7 mm proximally to the aneurysm and distally to the pulmonary arteries. Embolization of PAVM was successfully performed without residual shunt (fig. F).
Results: Postoperative period was uneventful, and patient was discharged on the 2nd postoperative day without any clinical signs. He was admitted after 3 months for examination. Boy was presented in the good state without symptoms, the right ventricular pressure was not elevated according to transthoracic echocardiography.
Conclusions: endovascular method of PAVM closure with occluder device is a safe and effective procedure. Amplatzer vascular plug has shown a good result of embolization.



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-203
Risk factors as predictors for an ASD-closure at very young age

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Background: According to current guidelines, an asymptomatic Atrial Septal Defect (ASD) is closed at the age of three to four years. Significant ASD will lead to volume overload, enlargement of the right side of the heart and several symptoms. The aim of this study was to assess risk factors for closure of ASD at two years of age or younger.
Methods: In this case-control study all children treated with ASD closure, surgically or with percutaneous device closure, between 2000-2014 at two of Sweden paediatric heart centres were included in the study. Cases were children at two years of age or younger at time of closure. Exposure information was retrieved from medical journals and national registries.
Results: Overall 413 children were included in the study population and 131 (32%) were two years or younger, equally distributed between ASD device closure and surgery. Risk factors associated with an early ASD closure were preterm birth, additional chromosomal abnormalities pulmonary hypertension and additional congenital heart defects, especially for an ASD size:body weight ratio of 0.8 and even after adjustments were made for confounding factors. An ASD size:body weight ratio of 2.0 as well as a ratio of 0.8, was associated with increased risk of an early ASD closure.
Conclusions: Several independent risk factors were associated with an increased risk of an early ASD closure. An ASD size:less body-weight ratio=2 or a ratio=0.8 were both associated with increased risk of an early ASD closure, indicating that the ASD size:body weight ratio is a poor predictor for indications of ASD closure.

P-204
Predictors of outcomes in pulmonary atresia with intact ventricular septum

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Background: Pulmonary atresia with intact ventricular septum (PA-IVS) is a congenital heart disease with a spectrum of severity than can lead to univentricular strategy or biventricular normal physiology. Predicting outcomes during the neonatal period remains challenging.
Objectives: To identify the predictors of biventricular repair versus 1.5 ventricle repair in PA-IVS and the risk factors for early mortality.
Methods: We retrospectively reviewed all neonates with PA-IVS over a period of 30 years. We characterized the outcomes as univentricular heart (1V) without attempt to decompress the right ventricle, 1.5 ventricle (1.5V) and biventricular repair (2V). The outcomes were the final type of repair and death. Right ventricle morphological and functional characteristics were analysed and the cut-off values of each measurement to predict the type of repair was evaluated.
Results: 248 patients were identified. 49 entered the 1V path (median z-score of tricuspid valve (TV) -3.42) and 199 had an attempt to decompress the RV (median Z-score of TV -1). Of those, 143 had a tripartite RV and 56 a bipartite RV. 16/57 patients with bi-partite RV and who had RV decompression finally entered the 1V path (median z-score of TV -3.25). 183 had a RV decompression with need for additional pulmonary blood flow in 82 (median z-score of TV -1.2) or without additional procedure in 101 (median z-score of TV +0.46). The predictors of 2V repair vs. 1.5 V were Z-score of TV > -1 (OR 5.1, CI95% 1.7-15.1), and no need for additional pulmonary blood flow (OR 7.6; CI95% 3.4-16.7). Risk factors for mortality were Z-score of TV > +1 (OR 5.4;CI95% 1-35.6), and severe tricuspid regurgitation (OR 10.4; CI95% 2.4-41.7).
Conclusion: The size of the inlet of the RV estimated with the z-score of the TV predicts the type of repair in PA-IVS as well as the need for additional pulmonary blood flow after RV decompression. Patients with high z-score of the TV and more than moderate tricuspid regurgitation have the highest mortality. They should potentially have a rapid closure of their arterial duct after RV decompression to avoid reverse flow from the aorta to the right atrium.

P-205
Long term outcomes after percutaneous pulmonary valve implantation in complex right ventricular outflow tracts using the "folded" Melody® valve technique

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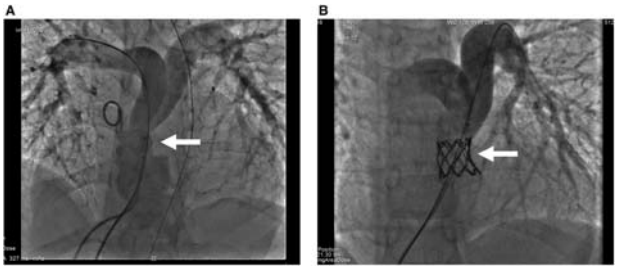
Introduction: Percutaneous pulmonary valve implantation (PPVI) has been validated as a valuable therapeutic option for the management of patients with dysfunctional right ventricular outflow tracts (RVOT). In these complex lesions, we reported that the implantation of a modified and "folded" Melody® valve was feasible and provided good early hemodynamic results
Methods: This is an observational multicenter study conducted in French tertiary Centers, between April 2012 and November 2018. Procedural and follow-up data of patients implanted with a "folded" Melody® valve were collected retrospectively from medical records in order to identify the clinical and hemodynamic mid-and long-term outcomes including: survival rate, clinical conditions, valvular function, need for reintervention, stent fracture and infective endocarditis.
Results: PPVI using the folded valve technique was performed in 10 patients. Mean age at PPVI was 22.8 years old (range 12 to 41). 60% were males. Indications for the folding valve were short RVOT and early bifurcation of pulmonary arteries in 6 patients and bioprosthetic valves in 4. No complication occurred during procedures. All patients had excellent hemodynamic results (mean post PPVI RV-PA gradient was 12.2 mmHg, 4 patients had trivial pulmonary regurgitation (PR) and the remaining had no PR). After a mean follow up of 30.6 months (range 10 to 66 months), mean echocardiography assessed RVOT peak velocity was 2.9m/s. Only one patient had trivial pulmonary regurgitation and two had mild to moderate pulmonary stenosis. No patient had reintervention. No valve dysfunction nor stent fractures were observed. No patient developed Endocarditis. Survival rate after 30.6 of follow-up was 100%.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



Conclusions: The "folded valve technique" is a safe modification of the Melody® valve which provides good long term results without increased rate of valve related complications.
Folded Melody® valve implantation in a patient diagnosed of Truncus arteriosus type 2A with short RVOT and early PA bifurcation. (A) Angiogram showing short RVOT with early PA bifurcation (white arrow) and free PR; (B) Angiogram after Folded valve implantation showing good valve function. Note the terminal stent struts on either sides of the Melody® valve that were folded over itself and the significant decreasing of device's length (white arrow).

P-206
Multiple coronary fistula closure in patient with pulmonary atresia and intact ventricular septum
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Introduction: Multiple coronary fistula are often diagnosed in patients with pulmonary atresia and intact ventricular septum (PA/IVS). Re-opening of the RVOT can cause significant impairment of coronary circulation what leads to myocardial infarction and consecutive heart failure in the early stage of life (RV-dependend coronary perfusion). In those patients univentricular palliation is chosen due to significant coronary fistula blood flow. We report on a patient with PA/IVS in whom closure of multiple coronary fistulas enabled to achieve biventricular circulation.
Methods: A 2,5-year-old boy with pulmonary atresia and intact ventricular septum and multiple coronary fistula and suprasystemic RV pressure was treated initially with Formula Stent implantation into the intra atrial septum and a BT-Shunt implantation after 2 unsuccessful attempts of ductus arteriosus stenting. The size of the right ventricle and tricuspid valve annulus were in the normal range so biventricular circulation seemed to be possible.
Results: At the age of 2 months the first three coronary fistulas (RV-RCA) were closed (one with ADO AS5/2 occluder and Coil 4/2, the other with ADO AS4/2 occluder and the third one with ADO AS3/2). At the age of 4 months the atretic pulmonary valve was perforated with the Baylis radiofrequency system and dilated with 4 mm balloons. After a month the next catheterization was scheduled and this time the fourth RV-RCA fistula was closed with a Covinien Microplug 5 and two RV-LCA fistulas were closed with an Amplatz plug 6-6 and an Amplatz Plug AS5-2 respectively. The RVOT was dilated with a 6 mm balloon. The last RV-LCA fistula was closed 8 months later with an Amplatzer Plug II 8/7 and the RVOT was dilated with an 8 mm balloon. One year later in echocardiography the RVSP was estimated at 40 mmHg and good ventricular function after occlusion of all coronary fistulas was observed.
Conclusion: Percutaneous closure of multiple coronary fistula is feasible and effective and may enable to avoid necessity to perform univentricular palliation in patients with PA/IVS with multiple coronary fistula. Of course this is only feasible if RV-dependent coronary circulation is absent.

P-207
Biodegradable metal stents in congenital heart diseases - a bail-out or a bridging solution?
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Background: Bioabsorbable magnesium scaffolds (MgS) have been shown to be effective in the adult coronary system. We analyzed their role in the treatment of vascular stenosis in children.
Method: Since 2016 fifteen MgS with a diameter of 3.5 mm were implanted in 9 infants and children aged 15 days to 7.6 years. Eight MgS were implanted in pulmonary venous re-stenoses (PVS), five in pulmonary arterial stenosis including one in-stent stenosis, one into a stenotic brachio-cephalic artery and one in a recurrent innominate vein thrombosis.
Results: All patients clinically improved after the implantation of a MgS. The MgS began to lose integrity at 4 to 6 weeks after implantation. The innominate vein thrombosed early, while all other vessels remained open. Two patients died after 4 and 12 weeks not related to the MgS. Re-stenoses of up to 50% diameter reduction occurred in the group with PVS and between 0% and 20% in the other lesions. Four patients received further interventions after the MgS had lost their function and redilations with larger balloon diameters of up to 6 mm were performed to follow growth. The rapamycin coverage of the MgS did not cause noticeable side effects.
Conclusion: The MgS can be used as a bridging solution to treat severe vascular stenosis in newborns and infants in different lesions. Restenosis can occur after degradation, but neither vessel growth nor further interventions are hindered by stent material. Larger diameters and prolonged degradation time may improve therapeutic options.

P-208
Transcatheter replacement of pulmonary valve with Venus P-Valve: a single center experience in 15 patients
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Objectives: To report the short term results of transcatheter replacement of pulmonary valve with the Venus P-Valve
Methods: During a 10-month period, 15 patients underwent attempted transcatheter replacement of pulmonary valve with a Venus P-Valve. Of the 15 patients aged 26±12 years, all had moderate-to-severe pulmonary regurgitation and significant right ventricular dilation. Fourteen had postoperative Tetralogy of Fallot and 1 had aortic valve stenosis status post Ross procedure. The diameter of Venus P-Valve selected was 2-4 mm larger than waist diameter of the sizing balloon. The length selected was similar to main pulmonary artery length. Aspirin 100mg and Plavix 37.5 mg was given to each patient for 6 months.
Results: The procedure was successful in all 15 patients. Eight patients underwent a 6-month follow-up magnetic resonance imaging study. There were significant reductions of right ventricular end-diastolic volume index (RVEDVi) and right ventricular end-systolic volume index (RVESVi) (168 ± 11 vs. 126 ± 9 ml/m2, and 93 ± 16 vs. 69 ± 8 ml/m2, respectively, $P < 0.01$). But there were no significant changes in RV ejection fractions

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

(44.9±10.0 vs. 44.8±6.0%). No major complications occurred. One patient developed tarry stool then aspirin was discontinued. After a mean follow-up period of 5.9 ± 3.4 months, symptomatic improvements were documented in 11 patients. The most recent echocardiography showed the implanted valves in good position and trivial pulmonary regurgitation in all 15 patients. Very mild paravalvular leak was found in 4 patients.
Conclusion: Transcatheter replacement of pulmonary valve with Venus P valve is safe and effective in patients with moderate-to-severe pulmonary regurgitation.

P-209
Nickel Allergic Reaction Post ASD Device Closure
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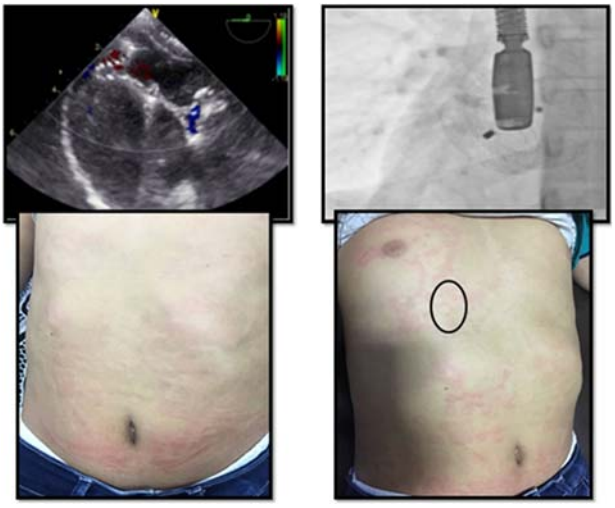
Introduction: The use of nitinol-containing devices can pre-dispose to nickel allergy. In 2009, a survey of CCISC determined the approach of interventional cardiologists to nickel allergy. Only 44% of responders routinely inquired about nickel allergy and performed skin testing prior to device closure. Reaction occurred from 2 days up to 1 month after implantation and manifested as headaches, rash/urticaria, dyspnea, fever or pericardial effusion. All patients responded to medical management and in rare instances the devices need to be explanted.
Methods: We report on 11 years old boy with dyspnea who diagnosed by TTE and TEE to have large secundum ASD measuring 29x27 mm with left to right shunt and insufficient flimsy posterior, superior and IVC rims. The right side showed dilatation (RVEDD=3.5cm) with normal PAP. The patient underwent successful transcatheter device closure in Tanta University catheterization laboratory using 34 mm Hyperion™ ASD Occluder (Comed) with no residual shunt or impingement over any of the cardiac structures. The patient was discharged on the same day on his previous medications (Spironolactone and ACEI) and on aspirin 3mg/kg/day oral and safely followed after one week.
Result: Ten days later, the patient complained of fever, severe allergic and pruritic reactions on his face, trunk and genital area. Echocardiography revealed no residual shunt or any complications. Aerobic and anaerobic blood cultures were negative and infective endocarditis was excluded.



After exclusion of infective endocarditis and drug reaction, there was a great concern of an allergy to the device itself. Cutaneous testing using 25 mm AMPLATZER™ PFO occluder was applied to the patient and showed positive results. Intravenous high dose dexamethasone was started immediately and tapered gradually over 5 days. Aspirin was replaced by clopidogrel by the same dose. Fortunately, two days later symptoms and signs started resolving gradually. The patient was discharged with safe and free 6 months follow up course.
Conclusion: Nickel allergy should be taken into account in patients considering an ASD device. Confirming a nickel allergy pre-procedure with patch testing or with the device itself is very useful procedure.

P-210
Catheter-interventional closure of bronchopleural fistula following pneumonia
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A bronchopleural fistula is a rare complication of pneumonia, lung abscessus or pleural empyema.
Casuistic: We report on an 2 ½ year old female infant who presented with a fever of 40°C, tachydyspnea of 70/min, marked respiratory distress, cough and back pain, in reduced condition on admission.
Clinically and radiologically a right sided pneumonia and pleural effusion were diagnosed. The child was started on antibiotics, non-invasive ventilation and received a pleural drainage. After removal of drainage 3 days after its cessation, a vavlular pneumothorax occurred. A new drainage was applied, but the air leak persisted for the next weeks. CT scan displayed collapsed right lung and septate pneumothorax. As surgical thoracoscopic debridement with new drainage remained unsuccesful, bronchoscopy was performed with bronchographic proof of a bronchopleural fistula in right-sided segment 3.
Intervention: After interdisciplinary consultation a bronchoscopy was performed with bronchography displaying the known bronchopleural fistula in segment 3 by application of contrast media. A Glidecath® 4 Fr and subsequent a microcatheter was placed in segment 3right. In the following superselective embolization with the use of Ruby Soft Coils® (2mmx1cm, 3mmx5cm, 4mmx6cm) and Ruby Standard Coil® 3mmx5cm was carried out followed by subsequent bronchographic proof of leak tightness. 2 days later, the drainage could be removed and the child was extubated. On follow-up over 2 months the coil position remained unchanged, and right sided chest x-ray controls normalized.
Discussion: A bronchpleural fistula is a very rare and severe complication of pneumonia or lung abscessus. On literature review, apart from surgical therapy by resection, coverage or peurodesis other interventional procedures like valve implantation, ASD-occluder device implantation, chemical intervention by topic administration of ethanol, silver nitrate, cyanoacrylate, fibrin glue, polyethylenglycol, doxycycline or cellulose have been published. However, in respect to the age and size of the child and very small anatomy, we decided for a to date non published interventional approach and closure by 4 Penumbra® coils. By this, a rapid removal of drainage and discharge was possible with uneventful controls over a 2 months' period. We can well recommend this alternative approach for lung air leakage due to bronchopleural fistula.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-211

Percutaneous pulmonary valve replacement in patients with left anterior descending coronary artery crossing the RVOT due to anomalous origin from the right coronary sinus

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Introduction: Percutaneous pulmonary valve implantation (PPVI) has found widespread acceptance in the postoperative revalvulation of the right ventricular outflow tract. Major obstacles for PPVI are coronary arteries in close proximity to the expected landing zone of the implanted valve. In patients with origin of the left anterior descending coronary artery (LAD) from the right coronary sinus the LAD crosses the RVOT and frequently precludes PPVI. We report our experience with PPVI in 4 patients with this coronary anomaly.
Patients: In our institution 66 patients underwent PPVI from 1/2010 – 11/2018. Origin of the LAD from the right coronary sinus was present in 4 patients (m=26,8 years). Underlying diagnoses were tetralogy of Fallot (2 pts.), PA-VSD and DORV. The morphology of the RVOT included s/p implantation of 18mm Contegra conduit, s/p 22mm homograft, s/p 18mm Goretex tube in addition to the native RVOT and RVOT following pulmonary valvotomy.
Results: The coronary arteries were displayed by selective injections and revealed the LAD crossing the RVOT at or just below the former pulmonary valve level. Balloon sizing was not performed in 2 patients with distal landing zones and calcified grafts. In the remaining patients balloon sizing was essential to define a landing zone distal to the LAD. In two patients the landing zone was short limiting the implantation to 26mm and 22mm Stents (LDmax and CP). In these patients the melody valve was shortened by folding of the proximal and distal struts. The third patient underwent implantation of melody valves both in the 18mm Goretex tube and in the native RVOT while the fourth patient underwent implantation of a 26mm Edwards Sapien valve. Selective angiographies following prestenting and valve implantation revealed unimpeded flow to the coronary arteries. The postinterventional course was uneventful and patients are doing well with a mean follow-up of 2,8 years.
Conclusions: According to our results anomalous origin of the LAD does not always preclude PPVI. The decision can be made on an individual basis depending on absence of proximal obstruction of the RVOT and on the possibility to create a landing zone distal to the LAD crossing the RVOT.

P-212

Validation of three-dimensional rotational angiography measurements in children with an aortic coarctation

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Introduction: Percutaneous interventions are increasingly used for treatment of aortic coarctation (CoA) in children. Successful intervention and follow-up requires adequate imaging of the cardiac and vessel anatomy and surrounding structures. Three-dimensional rotational angiography (3DRA) is a relative new technology to achieve this imaging. The aim of this study was to compare 3DRA aortic diameter measurements in children with CoA to computed tomography (CT) and magnetic resonance imaging (MRI) measurements.
Methods: Patients with CoA who underwent 3DRA guided percutaneous balloon angioplasty (BA) or stent implantations between January 2011 and March 2017 were retrospectively included. Aortic diameters were measured in a standardised fashion on 3DRA, CT and MRI.
Results: Sixteen patients that underwent 17 3DRA guided catheterisations were analysed, with a median age of 12.70 years (range 0.14-18.61 years) and a median weight of 51.50 kg (range 3.78-71.00 kg). CT as well as MRI measurements were significantly correlated with 3DRA measurements (Pearson $r = 0.679$, $p < 0.05$ resp. $r = 0.722$, $p < 0.05$). A mean underestimation of -0.77 mm (95%-CI -7.00 to 5.45, $p > 0.05$) for 3DRA was found compared to CT measurements. 3DRA compared to MRI measurements showed a mean underestimation in 3DRA of -1.59 mm (95%-CI -7.50 to 4.32, $p > 0.05$). Stent measurements gave primarily overestimation, whereas measurements after brachiocephalic artery branching showed primarily underestimation. The linear regression coefficient of the Bland-Altman plot was 0.067 ($p > 0.05$) and -0.052 ($p > 0.05$) for CT resp. MRI comparative measurements.
Conclusions: There is a mean underestimation of aortic diameters in 3DRA measurements compared to CT and MRI measurements, however, it was not statistically significant and large limits of agreement have been observed. The difference in measured aortic diameters varies with location of the measurement.

P-213

Three-Dimensional Rotational Angiography Guided Stenting to Optimize Pulmonary Blood Flow in Children with Single Ventricle Physiology

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Introduction: Stenosis of the Fontan circulation, in particular the pulmonary arteries (PA), is common in children with single ventricle physiology. Stent placement is the most used interventional strategy to optimize PA flow and Fontan hemodynamics.
Methods: A retrospective study was performed to investigate the prevalence, causes and outcome of Fontan circulation stenosis in children with single ventricle physiology treated with three dimensional rotational angiography (3DRA) guided stenting in our center.
Results: From September 2011 to October 2018 39 patients received 60 stents during 55 3DRA stent procedures. Median age and weight were 3.9 years (0.0-17.2) and 16.1 kilograms (4.1-70.0), respectively. Left pulmonary artery (LPA) stenosis accounted for 75% of the stenosis. In 22 cases the LPA stenosis was caused by external compression of the close interaction between ascending neo-aorta, descending aorta, pulmonary artery and airway. EV3 Mega/Max LD (N=33) and Cook Formula stents (N=21) were mostly used. Adverse events occurred in 5 catheterizations (10.6%). Fifteen patients (36.8%) underwent 19 reinterventions including planned serial redilation (N=12), extra stent placement (N=4) and surgery (N=1), and unplanned balloon dilation (N=1) and stent placement (N=1) for restenosis by intima proliferation.
Conclusion: Fontan stenosis, especially LPA stenosis, is common in children with single ventricle physiology and external compression is one of the main causes. 3DRA guided stenting is effective in both short and long term and can safely be performed in small children. Restenosis by intima proliferation is rare and may be

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

the result of the stent types used. Repeated redilations are obviously necessary to match stent diameter with patient growth.

P-214
Mid-aortic syndrome in childhood: case series

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Introduction: Mid-aortic syndrome (MAS) is defined as the stenosis of the abdominal or descending thoracic aorta and can be accompanied by narrowing of the visceral arteries (up to 66% of MAS involve renal arteries).MAS is an infrequent pathology, as it constitutes only 0.5-2% of all the coarctation of the aorta. The etiology of this disease is unknown and most of the cases are idiopathic, but some of them can be genetic or secondary to vasculitis such as Takayasu's arteritis. In childhood, it is typically presented as severe arterial hypertension.
Management of these patients is still unclear, as the treatment can be pharmacological or, in severe cases, surgical or endovascular.
Methods: Retrospective review of four cases of MAS in a university hospital.
Results: Four patients diagnosed with MAS were included in the review, with a mean age of 11.3 years (6-14 years old),3 of them were women. All the children were referred to our centre after the discovery of asymptomatic arterial hypertension, while MAS was diagnosed by magnetic resonance angiography in 3 of them and computed tomography angiography in the other patient.
Regarding the location of the lesions, one patient had stenosis of the suprarenal aorta, 2 of them of the paravisceral aorta and one of the descending thoracic aorta. Two of the children also had involvement of the visceral arteries: one of them, the celiac and both renal arteries, and the other patient the celiac, superior mesenteric and both renal arteries.
Endovascular treatment with percutaneous angioplasty and stenting was performed in 2 patients with good angiographic result and short-term arterial hypertension control. Although, one of them required two more angioplasties at 13 and 27 months due to refractory arterial hypertension.
The youngest patient was managed conservative with medical treatment with normalization of blood pressure. The remaining patient is waiting for endovascular treatment.
Conclusions: MAS is and infrequent disease with complex management due to the lack of scientific evidence. Invasive procedures are recommended in patients with severe arterial hypertension with end-organ damage or failure of the medical treatment. In those cases, endovascular procedures may be a safe option with acceptable results.

P-215
Safety and Efficacy of Palliative Stent Implantation in Premature Infants Under 1.4 kg

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Introduction: Premature infants with critical congenital heart disease pose difficult therapeutic problems: Surgery is associated with higher mortality and morbidity. Prolonged medical therapy to delay surgery does not improve survival. Palliative stent implantation may be an alternative in aortic coarctation and Tetralogy of Fallot.
Methods: To assess safety and efficacy of this approach, we collected demographics, procedural parameters, results, complications and late clinical outcomes of all premature infants <1.4 kg undergoing attempted palliative transcatheter stent implantation.
Results: Interventions were attempted in 8 patients (gestational age 26-34 weeks, median 30.5 weeks) and accomplished in all. Five patients (weight 900-1230 g; median 1050 g) underwent stent placement for coarctation with access by carotid cut-down. Fluoroscopy times were 4.4-10.8 min (median 6.6 min). The premounted stents were 3–4 mm in diameter and 7-9 mm in length. There were no procedural complications. One patient with prior septic episodes died from sepsis one week after the procedure; 3 had elective end-to-end repair after 4-5 months; 1 underwent Norwood after 3 months. Three patients underwent palliative stent placement for Tetralogy of Fallot (weight 970-1300g) by jugular vein cut-down. The premounted stents were 3.5–4 mm in diameter and 9-12 mm in length. Fluoroscopy times were 10.8–17.2 min. There were no complications. Elective repair was performed in all patients after 6–10 weeks.
Conclusion: Stent implantation appears to offer safe and effective palliation for premature infants allowing elective surgical repair at the preferred age and size.

P-216
25-year experience in percutaneous treatment of patent ductus arteriosus with different types of devices

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Introduction: Transcatheter closure has become a method of choice in treatment of patent ductus arteriosus (PDA). Different types of occluders have recently evolved. We present our own experience in this field.
Methods: Retrospective review included procedural data and one-year follow-up of all 974 patients (pts, 64.9% females) in whom transcatheter PDA closure was attempted between September 1993 and September 2018 at a tertiary referral centre. Median age and weight were 4.2 years (0.3-84.5) and 17.5 kg (3.9-136). We applied Rashkind device in 25 pts (R; 2.6%), detachable coil in 464 pts (C; 47.6%, multiple in 10 pts), Amplatzer duct occluder: type I or type I-like in 287 pts (ADO I; 29.5%), type II in 26 pts (ADO II; 2.7%) and type II additional sizes in 156 pts (ADO II AS; 16%). Other devices were used in special cases (16 pts; 1.6%).
Results: In PDA type A mainly ADO I were used and in types D and E coils, which were replaced with ADO II AS after introduction in 2014. Success rates were 88% for R, 96.8% for C, 96% for ADO I, 100% for ADO II and 99.4% for ADO II AS. There were 1 embolization in R group (4%), 7 in C (1.5%) and 1 case of iatrogenic coarctation of aorta with ADO II. Complete occlusion rate at 24 hours and a year after were, respectively, 77.3 and 86.4% for R, 83.3 and 91.8% for C, 96.5 and 100% for ADO I, 90.5 and 100% for ADO II and 98.1 and 100% for ADO II AS. 3 pts with R (13.6%) and 15 pts with C (3.3%) with residual shunt needed reintervention with coils. Amplatzer/Starflex septal occluders in PDA type B (n=7), muscular ventricular occluders in pts with pulmonary hypertension (n=6) and vascular plugs in PDA type D (n=3) were used with good clinical outcome. Fluoroscopy time was significantly shorter in ADO II AS (p=.000).
Conclusions: During last 25 years percutaneous treatment of PDA has significantly improved due to devices' development and diversity. Nowadays they are safe and efficient with high complete occlusion rate in all PDA types.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-217
Mortality and major adverse event related factors in Fallot Tetralogy

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Introduction: In this study, we aimed to determine the risk factors associated with mortality, long hospitalization and complicated postoperative period in patients who underwent Fallot tetralogy (TOF) complete correction.
Methods: A total of 170 consecutive patients, who were operated between January 2014 and June 2018 were retrospectively reviewed. TOF-Pulmonary valve absence and TOF-Pulmonary atresia (PA) were not included in the study. Surgical results and complications were determined according to international standards. Significant residual lesion necessitating reoperation, permanent pacemaker placement due to complete atrioventricular block (AVB), acute renal failure, need for ECMO support, neurological event and death were defined as major adverse events (MAE). Prolonged intensive care unit (ICU) stay was defined as more than 3 days and prolonged hospital stay was defined as more than 7 days.
Results: The mean age was 12 (1-192) months. Palliative procedures were performed in 26(15.2%) patients and primary repair was performed in the remaining 144(84.8%) patients. 115(67.6%) patients had transannular patch (TAP) and 35(20.5%) of these had additional anterior leaflet augmentation, 41 (24.3%) patients had ventriculotomy with non TAP repair, 10(5.9%) had transpulmonary + transatrial repair and 4(2.4%) patients had RV-PA conduit replacement. The mean postoperative RV / LV pressure ratio was 0.5 (0.2-0.7). AVB was seen in 5(3.0%), neurological events seen in 3(1.8%) patients. ECMO support was needed in 7 patients (4.1%). The overall in-hospital mortality was 3.5%. Small annulus z score (p=0.01), significant VSD after operation (residual or additional VSD) were risk factor for mortality (p=0.03) and MAE (p=0.02). High preoperative hematocrit level prolonged hospital stay (p=0.001) (Table 1).
Conclusions: Elevated preoperative hematocrit level due to desaturation, as a sign of longer preoperative period, is related to prolonged hospital stay and higher cost. We observed that residual or additional VSDs after TOF total repair were associated with mortality and major adverse events. Residual or additional VSDs should be closed surgically or percutaneously if possible.

Table 1: Predictors for Mortality, Major Advers Events, Lenght of Hospital Stay

	Predictor	OR	95% CI lower	95% CI upper	P value
Mortality	Annulus z score	0.5	0.3	0.9	0.01
	Residuel or additional VSD (2-4 mm)	54.6	1.6	1874.2	0.03
Majos Advers Events	Residuel or additional VSD (2-4mm)	12.4	1.5	99.9	0.02
Lenght of Hospital Stay	Preoperative Hematocrit	1.12	1.1	1.2	<0.001

P-218
Aortic and pulmonary valve replacement with the Inspiris Resilia valve in congenital heart disease

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Introduction: Valve replacement in young adults or pediatric patients is a complex decision and the kind of prosthesis implanted should be carefully selected.
Inspiris Resilia valve (Carpentier-Edwards) is a new bovine bioprosthesis that combines:

1) RESILIA tissue, a technology with anti-calcification properties, improved hemodynamic performance and dry storage.
2) VFit technology, an expandable frame designed for potential future valve-in-valve procedures.

The objective of our study is to report the results with this valve in patients with congenital heart disease.
Methods: We performed a retrospective observational study. Demographic, echocardiographic and follow up perioperative and postoperative data was recorded from our hospital database in patients with congenital heart disease that received an Inspiris Resilia valve since December 2017 to November 2018.
Results: 12 Resilia Inspiris bioprosthesis were implanted; 6 (50%) in pulmonary position in patients with a tetralogy of Fallot and severe pulmonary regurgitation and 6 (50%) in aortic position in congenital bicuspid aortic valves.75% of the patients were male, with a mean age of 37±18 years and 3 patients (25%) were under 18 years old. Valve size was 21 in 3 case(25%), 23 in 6 patients (50%) and 3 patients received a 25mm valve (25%). There were no mortality and all patients were discharged home without complications. For a mean follow up of 7, 38 months we don´t find any clinical complication and all prosthesis remains with a normal function in echocardiogram, with a mean peak across the valve of 23.75 ±10.70 mmHg.
Conclusions: In our experience, the implant of Inspiris Resilia valve either in pulmonary or aortic position in patients with congenital heart disease shows excellent short term results, similar to other bioprosthesis. A longer follow up is needed to evaluate mid and long term results, although we consider this valve as a good option in this population not only for the durability but also for a promising future in the valve in valve procedure.

P-220
Evaluation of Postoperative Respiratory System Complications in Neonates Operated for Congenital Heart Disease

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Objective: Despite advances in surgical technique and monitorization, postoperative complications in newborns undergoing cardiac surgery are important cause of morbidity and mortality even in developed countries. Respiratory complications some of which are preventable are among the most significant ones and may adversely affect the outcome. We aimed to investigate retrospectively the incidence of respiratory complications in newborns after cardiac surgery, risk factors for occurence and effect on morbidity and mortality.
Methods: A total of 198 patients under 1 month of age who underwent cardiac surgery between January 2011 and January 2016 were evaluated retrospectively. Gender, birth weight, weight at the time of operation, cardiac diagnosis, incision type, surgery, operation time, aortic clamp time, cardiopulmonary bypass time, duration of stay in intensive care unit (ICU), mechanical ventilation,

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

requirement of reintubation, postoperative complications of the respiratory system, other complications in the intensive care unit, mortality and presence of arrhythmia were recorded. Operation notes, ICU data of the patients were examined. Effect of respiratory complications on morbidity and mortality is investigated. Statistical significance level was accepted as $p < 0.05$.
Results: Postoperative respiratory complications developed in 92 (46.5%) of the 198 patients (131 were male, 61.7%). Diaphragmatic paralysis occurred in 29.3% of those with respiratory system complication, prolonged pleural effusion in 25%, pneumothorax in 14.1% and atelectasis in 12%, lung infection in 6.5%, vocal cord paralysis in 3.3%, chylothorax in 3.3%, subglottic stenosis in 3.3%, subglottic stenosis with vocal cord paralysis in 1.1%, subglottic stenosis with pneumothorax in 1.1% and finally subglottic stenosis with diaphragmatic paralysis in 1.1%. We found that diapragmatic paralysis was the most common respiratory complication.
The duration of mechanical ventilation, duration of stay in ICU, duration of stay in hospital were all longer in patients with respiratory complications than those without.
Conclusion: The presence of respiratory system complications such as subglottic stenosis, diaphragmatic paralysis, prolonged pleural effusion, pneumothorax, pulmonary infection causes prolonged stay in ICU and increases risk of reintubation in neonates who underwent cardiac surgery.

P-222
Whether the proposed of new pulmonary hypertension criteria has an impact on pediatric and adolescent patients?

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According to the 6th World Symposium on Pulmonary Hypertension, there was a new proposed criterion for pulmonary hypertension (PH) diagnosis with the mean pulmonary artery pressure (mPAP) $\geq$ 20 mmHg instead of $\geq$ 25 mmHg. Very little data is available for pediatric and adolescent pulmonary hypertension patients whether the diagnostic criteria change would have an impact of patient care.
Objective: To define whether lower the bar for pulmonary hypertension diagnosis would increase the number of pediatric and adolescent patients and change their management.
Method: Retrospectively review of the previous cardiac catheterization record, the data was sourced out and defined the PH cases base on the existing criteria versus the newly proposed criteria. The patients were divided into 3 groups according to their mPAP, specifically group A: < 20 mmHg, group B: 20-24mmHg, and group C: 25mmHg.
Result: A total of 85 patients underwent cardiac catheterization for hemodynamic evaluation with complete data record at a tertiary care center specialized in pediatric pulmonary hypertension. Group A composed of 32 patients with mean age of 8.95 years and mean mPAP15 mmHg. There were 8 patients in group B with their mean age of 10.7 years and mean mPAP 21.7 mmHg while their pulmonary vascular resistance < 3 WU.m2. While 44 patients in group C, mean age 7.41 years, had their mean mPAP 28 mmHg.
Conclusion: With the newly proposed PH criteria, there was 18% increase in number of patients diagnosed with PH in comparison with previous criteria for PH diagnosis. This group of patients, who had mPAP 20-24 mmHg, had pulmonary vascular resistance less than 3 WU.m2. Therefore, the increase number of patients, due to newly criteria for PH diagnosis, did not have a significant impact on patient management in term surgery management or pulmo.

P-223
Utility of the Inotropic and Vasoactive Score in the Analysis of Morbidity and Mortality in Pediatric and Neonatal Cardiac Surgery

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Objective: To describe the morbidity and mortality associated with pediatric and neonatal cardiac surgery and to analyze the usefulness of the Vasoactive-Inotropic Score (VIS).
Material and methods: Retrospective and descriptive study. Patients undergoing cardiac surgery in a period of 19 consecutive months (2017-2018). A database with demographic, surgical and postoperative management variables was used. Surgeries were classified according to the STAT category [high (4-5) vs low (1-3)] and VIS (high ($\geq$20) vs low (<20)].
Results: We analyzed 335 consecutive surgeries, 261 (78%) with extracorporeal circulation, in patients aged between 1 day and 18 years [91 (27.2%) neonates and 244 (72.8%) pediatric]. Sixty-eight (20.3%) were high STAT. 81.2% of the patients were extubated before 48 hours. The most used vasoactive drugs were milrinone and dopamine. Thirty-five (10.4%) had high VIS.
In pediatric patients, the ICU stay was longer in surgeries with high STAT [6 days (2-8,5)] vs low STAT [3 days (2-5)], ($p = 0.08$); and in high VIS [16 days (7-23)] vs low VIS [3 days (2-5)], ($p < 0.01$). In the neonates, no significant differences were observed, high STAT [17 days (7-31)] vs low STAT [9 days (7-20,75)], ($p=0.06$); and high VIS [17 days (8,25-33,25)] vs low VIS [12 days (7-25)], ($p=0.14$).Need for ECMO was reported in 5(1.8%) patients, renal replacement therapy in 7(2%) and diaphragmatic plication in 10 (2.8%). Nineteen patients were re-operated. Hospital mortality was 0.89%.
Conclusion: Our center has a low mortality rate in pediatric cardiac surgery. In pediatric patients, high STAT category and elevated VIS are associated with longer hospital stays. However, these differences are not observed in the neonatal population. Future studies should focus on optimizing the categorization of risk and surgical morbidity in neonatal patients.

P-224
Flail of the tricuspid valve as a manifestation of neonatal lupus

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Introduction: (or Basis or Objectives): Valve dysfunction is not a well-known feature for infants of anti-Ro/SSA-positive pregnancies, but anecdotal cases have suggested the association between rupture of the AV valve tensor apparatus and maternal anti-Ro/SSA antibodies.
Methods: We present a case of a patient with valve dysfunction secondary to rupture of the papillary muscle of tricuspid valve affected by maternal anti-Ro antibodies.
Results: Routine 20 week gestation fetal echocardiography of a first gravida mother revealed focal areas of increased echogenicity at the level of the papillary muscles and chordae of both AV valves (Figure). Heart rhythm was normal. During the following weeks those patchy areas disappeared. Neither insufficiency nor stenosis

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

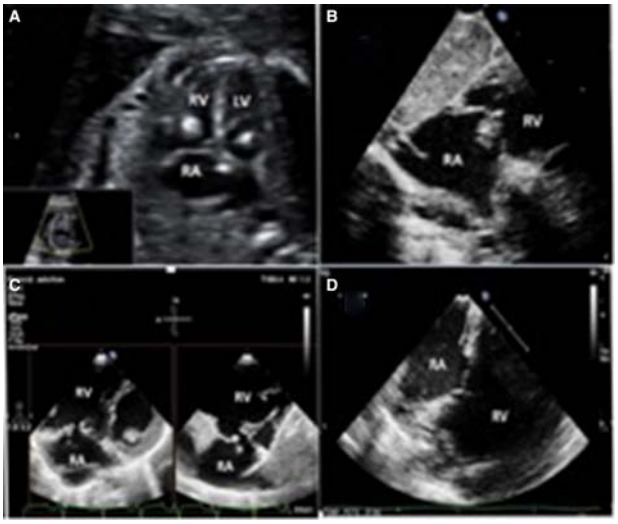

Figure.
Fetal echocardiogram (a) with localized areas of increased echogenicity at the papillary muscles of both AV valves. Postnatal echocardiogram showed severe insufficiency of the tricuspid valve and an image that reminded a vegetation (b). Echocardiogram showed a flail of the tricuspid valve with severe regurgitation and dilatation of the right ventricular cavities (c). Postoperative results with good leaflet coaptation and mild regurgitation (d).

was detected. Rheumatologic prenatal profile was positive for anti-Ro/SSA antibodies and anti-La/SSB without clinical systemic lupus erythematosus and other connective tissue. At 39 weeks Emergency caesarean section was performed due to fetal hydrops. After birth, the patient required high-frequency ventilation, nitric oxide, inotropic support and diuretic.Postnatal echocardiography showed normal biventricular function and severe tricuspid regurgitation, prolapse of the anterior leaflet with and image that suggested a vegetation (Figure). Laboratory study was positive for factor V Leiden mutation and anti-Ro/SSA antibodies; anti-La/SSB was negative.
Transesophageal echocardiography revealed a flail of the anterior tricuspid valve leaflet. Intraoperative findings showed disruption of the chordal attachments of the anterior leaflet without the presence of a papillary muscle. From the right ventricle a thick bundle muscle of the free wall was detached and used as a neo-papillary muscle. The anterior leaflet of the tricuspid valve was directly attached to the neo-papillary muscle with prolene 5/0. A tricuspid annuloplasty was also realized (Figure). Biopsy specimens from the right ventricle were obtained.
Conclusions: The pathogenesis of the papillary muscle chordal rupture may be similar to that proposed for autoimmune AV block, binding in this case the antibodies to the cardiac myocytes and evoking an inflammatory response with subsequent fibrosis and ultimately muscle chordal rupture.

P-225
Incidence and spectrum of congenital heart disease in the neonatal intensive care unit at high altitude in China: A retrospective study

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Introduction: Previous studies including ours screening congenital heart disease (CHD) at high altitudes have reported substantially higher incidence as compared to that at low altitude, consisting almost solely of simple CHD with left to right shunt. Data on the occurrence of complex CHD at high altitude is scarce. Neonates with complex CHD are likely to be admitted to NICU. Therefore, the present study aimed to evaluate the incidence and spectrum of CHD in NICU in order to depict a truer picture of CHD at high altitude.
Methods: We retrospectively reviewed charts of 4214 neonates in the Women's and Children's Hospital in Xining (2,260 m), Qinghai province (average altitude 3000 m) in 2015-2016. Echocardiography was performed in 1943 babies (aged 10 minutes to 2 months; 1220 boys, 723 girls; altitude ranged 1,800 to 4,300 m, median 2,526 m) when CHD was suspected based on heart murmur, cyanosis or pneumonia.
Results: CHD was diagnosed in 1093 babies (56.3% in echoed 1943 babies), making the incidence in total NICU patients 26%. They were hospitalized mainly because of pneumonia (62.6%), asphyxia (13.2%) and hyperbilirubinemia (7.3%). Mild and moderate CHD accounted for 97.6%(1067 babies), including 583 (53.3%) secundum atrial septal defect, 227(20.7%) patent ductus arteriosus, 20(1.8%) ventricular septal defect, 263(24.1%) multiple defects with left to right shunt, 1(0.1%) bicuspid aortic valve, 7 (0.6%) pulmonary stenosis, 2(0.2%) aortic stenosis. Severe CHD accounted for 2.4% (26), including 6(0.5%) atrioventricular septal defect, 5(0.5%) complete transposition of the great arteries (TGA), 6(0.5%) hypoplastic right heart, 3(0.3%) hypoplastic left heart, 3 (0.3%) double outlet right ventricle, 3(0.3%) tetralogy of Fallot, 2(0.2%) truncus arteriosus, 2(0.2%) total anomalous pulmonary venous connection, 2(0.2%) severe aortic stenosis, 2(0.2%) severe pulmonic stenosis. In 26 patients with severe CHD during the period of 2 to 12 months after discharge, 17 patients died before cardiac surgery, and 1 of the 4 survivors had arterial switch operation, and 5 lost track.
Conclusion: Our study provides the initial information about the wide spectrum of complex CHD at high altitude, This combined with high mortality, indicates the urgent need for the implementation of routine echocardiography at NICU and follow-up program in high altitude regions.

P-226
Epidemiology of Acute Kidney Injury Among Pediatric Patients with Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery after Repair over 7 Years

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Introduction: Acute kidney injury (AKI) presents as a prevalent complication after surgical repair of pediatric cardiac deficiency and is associated with poor outcomes. The state of insufficient renal perfusion secondary to severe myocardial dysfunction after birth is probably an independent risk factor in patients undergoing anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) repair. We retrospectively figure out the epidemiology of pediatric acute renal outcome in ALCAPA population after repair
Methods: There were a total of 89 children included who underwent left coronary reimplantation. Pediatric-modified risk, injury,

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

failure, loss and end-stage kidney stage (pRIFLE) criteria was chosen to make the diagnosis of AKI.
Results: The incidence of AKI was 67.4% (60/89) in our study. Among AKI cohort, 23 (38.3%) patients were diagnosed as AKI-I/F (20 in I criteria and 3 in F criteria). The poor cardiac dysfunction (Left ventricular ejection fraction (LVEF) less than 35%) prior to surgery was a significant contributing factor associated with onset of AKI (OR, 5.553, 95% CI, 1.393-22.132; p = 0.015), while longer duration of anomaly discovery until surgical repair (OR, 0.973, 95% CI, 0.946-1.000; p = 0.049) and preoperative higher albumin level (OR, 0.831, 95% CI, 0.696-0.992; p = 0.041) were found to alleviate AKI condition. Neither preoperative severity of mitral regurgitation nor mitral annuloplasty was associated with AKI onset. After reimplantation process, there was 1 death in no-AKI group and 2 deaths in AKI-I/F (p = 0.356), and other children all have survived until hospital discharge. Median follow-up time was 46.5 months (IQR, 34.0-63.25 months). And during their follow-up time, patients in AKI cohort were seen more by specialists and rechecked more by echocardiography.
Conclusions: Pediatric AKI after ALCAPA repair occurs in a relatively higher incidence than previous cardiac reports and was linked to clinical outcomes. Preoperative poor cardiac dysfunction (LVEF < 35%) is strongly associated with AKI development. Moderately prolonging the duration from coronary anomaly discovery till surgical repair is needed to take medical measures to optimize the decreased cardiac function and the poor nutrition in terms of decreasing acute kidney problem.

P-227
Cardiopulmonary bypass is associated with insulin resistance and inflammation in congenital heart disease with or without increased pulmonary blood flow

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Objective: Cardiopulmonary bypass (CBP) is associated with inflammation and altered glucose metabolism. Increased pulmonary blood flow (PBF) has been shown to affect circulating and tissue cytokine levels in children with left-to-right cardiovascular shunt lesions.
Aim: To analyze blood plasma concentrations of proinflammatory cytokines, insulin, and insulin-sensitizer adiponectin in infants with congenital heart disease (CHD) pre and post CPB. To assess whether cytokine levels are correlated with low cardiac output syndrome (LCOS) after CBP.
Methods: We conducted a prospective study including 58 infants, who underwent CBP (normal or decreased PBF: ToF n=19; increased PBF: VSD n=24, AVSD n=9; single ventricle: HLHS n=6). Plasma cytokine, insulin and adiponectin levels were measured by RIA and ELISA.
Results: There were no significant differences in the plasma levels of TNF-alpha, IL-6, IL-10, MCP-1, and RANTES between the 4 CHD groups, before and after CPB. IL-6 levels increased after CPB in all groups, while TNF-alpha and RANTES decreased after CPB in ToF and VSD. IL-10 and MCP-1 expression did not change after CPB. Postoperative LCOS was associated with higher MCP-1 and lower RANTES levels before and after CPB, and higher TNF-alpha levels after CPB. Insulin levels before CPB were significantly higher (5-8 fold) in HLHS patients compared to all other groups. After CPB, insulin increased in ToF and VSD patients, while adiponectin decreased by 30-60% in all 4 groups. Patients with VSD or AVSD had the highest plasma adiponectin levels, both before and after CPB.
Conclusion: CPB is associated with inflammation and insulin resistance. A differential immunomodulatory plasma cytokine pattern exists in children with LCOS after CPB (MCP-1 and TNF-alpha increased, RANTES decreased). While infants with VSD or AVSD (i.e., increased PBF) have higher vasoprotective adiponectin levels after CPB, proinflammatory markers do not correlate with PBF.

P-228
Technical Performance Score in a single center in Argentina

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Objectives: Technical Performance Score (TPS) was developed as a tool to evaluate surgical repair in certain procedures, and proved to be useful in predicting early outcomes. The objective is to describe our surgical performance and results according this score.
Methods: consecutive patients with VSD, CAVC, TOF repair and ASO procedures from January 2016 to March 2018 were included. Demographic, surgical data, postoperative course and discharge were retrospectively reviewed. Surgical repair was considered: optimal, adequate or inadequate according echocardiographic criteria. Outcomes evaluated were early mortality, adverse events and postoperative length of stay. Adverse events recorded: extracorporeal membrane oxygenator support; re-exploration for bleeding, diaphragm plication or infection; cardiac arrest requiring resuscitation; stroke; and renal failure requiring dialysis. Unplanned reinterventions in the treated anatomic area and placement of permanent pacemakers were not included, because they are components of the TPS. Statistical analysis: categorical variables are summarized as numbers and percentages and continuous variables as medians and ranges. Chi square or Wilcoxon Rank Sum Test were used for differences in outcomes according TPS score.
Results: 258 patients were included, median age 141 days (1-5342) and weight 7 kg (3-53), and genetic syndrome association in 33%. Procedures distribution were 45% VSD closure, 20% TOF repair, 18% ASO, 16% CAVC repair. Average bypass was 102 min ± 40 and ACC 76 min ± 32. Assigned TPS was optimal in 38.7%, adequate in 54.2% and inadequate in 6.9% cases. Postoperative median length of stay(PLOS) was 6 days (1-160); 3.4% required renal replacement, 1.9% ECMO and 5.4% unplanned reoperation. Postoperative early mortality was 4.8%. Results grouped by TPS are describe in the table.
Conclusions: TPS was optimal or adequate in 93.1% surgical repairs. TPS class 3 was associated with higher percentage of adverse events and early mortality in our population.

	TPS1	TPS 2	TPS 3
PLOS days	6 (1-85)	6 (1-106)	12.5 (0-54)
Mechanical ventilation days	1 (1-29)	2 (1-90)	5 (1-23)
Mortality (%)	1	4.3	26.3
Adverse events (%)	5	6.4	26

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-229

Management of patent ductus arteriosus in preterm babies is not a surgical consideration and can be managed conservatively

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Introduction: Significant left to right shunt across the patent ductus arteriosus (PDA) in preterm babies is associated with higher incidence of intraventricular haemorrhage (IVH), necrotising enterocolitis (NEC) and chronic lung disease (CLD), and longer duration of ventilation. Surgical management has often been recommended as the final option. Although there are wide variations in medical management of PDA, the value of surgical PDA ligation or interventional device closure is not much questioned.
Objectives: This review looks at the clinical outcomes of premature babies with large PDA managed prior to 2014 (the era of surgical/medical intervention) versus conservative management, following a major shift in our practice in South Wales in 2014. New guidelines advocate liberal use of steroid, diuretics & fluid restriction, and limited use of Ibuprofen or Paracetamol.
Methods: Retrospective review of all preterm infants managed in our neonatal unit with a haemodynamically significant PDA. We studied outcomes of PDA, incidence of NEC, CLD, IVH, surgery/intervention and death.
Conclusions: Conservative management of PDA in preterm babies is as effective and results in fewer patients being referred for surgery or intervention without adversely affecting mortality or morbidity. Our study challenges the need for surgery or device closure in the management of preterm PDA.
Results:

	Medically treated		Not medically treated	
	Pre 2013	**2014-2017**	**Pre 2013**	**2014-2017**
N	88(44%)	20(18%)	113(56%)	89(82%)
Median gestation	26/40	25/40	27/40	27/40
Median BW (grams)	850	800	1020	955
Surgery/Cath	33 (37.5%)	1 (5%)	21 (18.5%)	2 (2%)
NEC	14 (15.9%)	6 (30%)	20 (17.6%)	25 (29%)
Death	12 (13.6%)	3 (15%)	13 (11.5%)	9 (10%)

P-230

Series of Cases of Coronary Anomaly with Origin in Pulmonary Artery in Children

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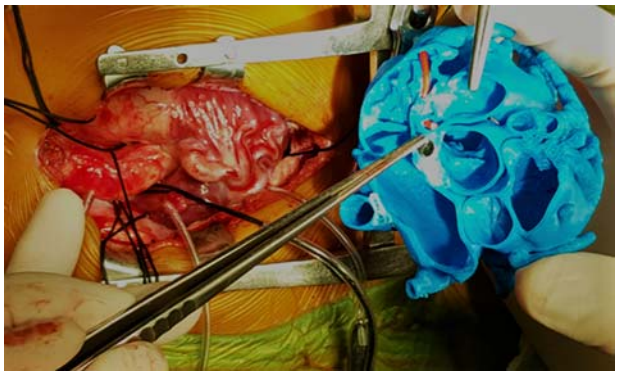
Introduction: Coronary artery anomalies occur in 1.3% of the population. The anomalous origin of the coronary artery from the pulmonary artery is a subtype of these anomalies (ACAPA). The left coronary artery anomaly (ALCAPA) occurs in 0.25-0.5% of congenital heart diseases and the right coronary artery (ARCAPA) occurs in 0.022%.Objectives: Presentation of a series of ACAPA cases in the last two years.
Methods: We retrospectively studied the cases of children from 1 month to 16 years with diagnosis and surgical treatment of ACAPA from 2016-2018 in Children's Hospital of the Santisima Trinidad de Córdoba.
Results: 4 patients, from 5 ms to 11 years of female sex (table 1)
Conclusions: Given the high mortality of patients with ACAPA in pediatrics and the current good surgical results (83%) (Walsh et al),

Table 1: Comparison of population and results of patients studied. Angiography: Provided data on collaterals and dilation of the coronary artery of normal origin. All patients underwent surgery with coronary reimplantation, with good results

	Patient 1	Patient 2	Patient 3	Patient 4
Age	5 mth	8 mth	8 mth	11 years
Diagnostic	ARCAPA	ALCAPA	ALCAPA	ALCAPA
Clinics findings	Asyntomatc	Asyntomatic	Heart faliure	Heart faliure
ECG	Normal	Normal	HLV	Subendocardal ischemia
Systolic funtion of LV	Normal	Normal	Severe dysfunction	Mild disfuntion
Echocardiogram			Mild MR and fibroeslatosis	Moderate MR y fibroelastosis
TC with 3D reconstructon	yes	yes	No	No



3D Anatomy de ALCAPA during the surgery

early diagnosis and high clinical-electro and echocardiographic suspicion are crucial for any patient with symptoms of irritability or dyspnea and continuous murmur or persistence of it after surgical correction of ductus, alterations in the ST and / or presence of heart failure accompanied by dilated cardiomyopathy

P-231

Cardiac insufficiency as a main predictor for persistent effusions after surgery on congenital heart disease

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Objectives: This study analyses different pre-, intra- and postoperative risk factors for chylothorax and persistent serous effusions (>7d) after congenital heart surgery and develops equations to calculate probabilities for their occurrence.
Methods: Retrospective review of different medical databases at the University Hospital of Erlangen between 01/14 and 12/16. Full model regression analysis was used to identify risk factors. Logistic equations were set up to calculate probabilities. Discriminative power of the developed models was checked with the c-statistics.
Results: Sixty-eight of 745 patients developed chylothorax (9.1%) and 125 of 677 persistent serous effusions (18.5%). Lower temperature (p=0.043; OR=0.899), Trisomy 21 (p=0.001; OR=5.548), a higher VIS at the day of surgery (p=0.001; OR=1.070) and assist device usage (p=0.001; OR=5.779) were significantly associated with chylothorax. Risk factors for persistent serous effusions were a given or possible involvement of the aortic arch during the operation (p=0.000; OR=3.982 und 2.905), univentricular hearts (p=0.019; OR=2.644), a higher number of previous heart surgeries (p=0.014; OR=1.436), a higher VIS at 72h after surgery (p=0.019; OR=1.091), a higher CVP at surgery (p=0.046;

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

OR=1.076)and an AoX time > 86min (p=0.023; OR=2.223), as well as assist device usage (p=0.002; OR=10.281). Both types of effusions were associated with a significantly higher morbidity and mortality.
Conclusion: Persistent serous effusions is associated with postoperative cardiac insufficiency, represented by a higher vasoactive ionotropic score at 72h after surgery, an AoX time > 86min and elevated CVP directly after surgery. The developed logistic equations help to estimate likelihoods in the future.

P-232

Severe over-anticoagulation despite standard phenprocoumon initiation protocol after aortic valve replacement in two paediatric patients

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Introduction: Vitamin K antagonists (VKA) are loaded routinely with standard clinically-based initiation protocols in children early after mechanical valve implantation, as this allows patients to reach target international normalised ratio (INR) rapidly without prolonged hospitalisation.
Cases: We present two boys aged 7 (patient 1 (P1), body weight 18 kg) and 16 years (patient 2 (P2), body weight 61 kg) who underwent mechanical aortic valve replacement. Postoperatively, overlapping with unfractionated heparin, we began oral anticoagulation with phenprocoumon using a standard body weight-based dosing algorithm. Target INR was 2.5 – 3.0. After only two doses of phenprocoumon (on day 1/day 2: P1 4.5 mg/1.5 mg, P2 9 mg/6 mg) both patients demonstrated an INR >7 in venous blood samples. Due to the significant risk of bleeding we administered vitamin K to both and fresh frozen plasma to P2. Hereafter, both patients required unusually low phenprocoumon doses of under 15% of normal (P1: 0.1 mg every three days, P2: 0.75 mg per day) to reach target INR. We performed sequencing of the vitamin K epoxide reductase complex 1 (VKORC1), cytochrome P450 2C9 (CYP2C9), and cytochrome P450 4F2 (CYP4F2) genes and found a homozygous VKORC1:c.-1639AA haplotype and a heterozygous CYP4F2 wild type (CYP4F2*1*1) haplotype in both patients, as well as a heterozygous CYP2C9*1*2 haplotype in P2. This confirmed enhanced sensitivity to VKA in both patients, leading to severely reduced phenprocoumon dose requirement.
Conclusions: Aside from age-related factors, genetic variations in VKORC1, CYP2C9, and CYP4F2 have been associated with significant inter-individual VKA dosing variability. Although infrequent, these variations may pose a significant risk for over-anticoagulation in children. Therefore, when using a standard clinically-driven loading protocol, patients with increased VKA sensitivity due to unknown mutations are at risk for bleeding. On the other hand, a low-dose initiation protocol would lead to prolonged hospitalisation in the majority of the children, who are unaffected by these genetic variations.
The routine application of a genotype-guided algorithm for phenprocoumon initiation therapy remains unrealistic in daily paediatric cardiac surgery. Therefore, it is even more crucial to be aware of this potential risk and to interpret the daily blood samples with care.

P-233

Norwood patients possess mild inferior cardio-pulmonary circulations even after Fontan procedure

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Introduction: We perform Norwood procedure (NWD) against patients with hypoplastic left heart syndrome. Post-NWD patients almost all possess unstable hemodynamic status with small PA before Glenn, even after Glenn. We predicted NWD patients, who developed the condition for Fontan, would still possess inferior cardio-pulmonary performances. We investigated cardio-pulmonary circulation in Fontan patients with a history of NWD.
Methods: The medical records of 156 Fontan patients were reviewed who underwent cardiac catheterization between 2010 and 2017. We divide them into 2 groups: patients who were performed Norwood procedure before Glenn (NWD: n=34) and patients who did not have history of Norwood (non-NWD: n=122). We compared cardio-pulmonary performances between NWD group and non-NWD group.
Results: We inserted conduit to pulmonary arteries on NWD and removed them on Fontan in all NWD patients. As to ventricular performances, ventricular volumes on end-systole were larger in NWD group than in non-NWD group (31 vs. 26 ml/m2: p=0.036). Ventricular ejection fractions were lower in NWD (52% vs. 56%: p=0.023). With regard to pressure studies, The patients rate with higher pressures of pulmonary capillary wedge (≥ 8 mmHg) was higher in Nor group (25% vs. 6%: p=0.025). There were no significant differences in other cardiac performances between two groups. As for pulmonary circulation, the ratio of patients with smaller pulmonary indexes (<145 mm/m2) was higher in NWD (25% vs. 6%: p=0.012). However, the ratio of patients was higher in NWD group (61% vs. 31%: p=0.0011) who undertook percutaneous transcatheter angioplasties. Pressures of superior vena cava were almost same between two groups; pulmonary resistances were not significantly different. Significant differences in the frequency of internal remedies did not existed between NWD and non-NWD, such as diuretics, vasodilators, beta antagonist, and pulmonary vasodilators. For enarapril and carvedilol, dosage amounts were not significantly different between two groups.
Discussion: Our study showed traces of unstable cardio-pulmonary circulation subsisted in NWD group even after Fontan procedure, especially cardiac systolic hypo-function and narrow pulmonary artery size. However, extent of internal medicines was not different at all between NWD and non-NWD. Patients after Norwood procedure continuously need intense medical management after Fontan procedure.

P-234

Risk of Thrombus formation and aortic regurgitation after repair of congenital coronary artery fistulas

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Introduction: Coronary artery fistula (CAF) is a rare congenital anomaly of coronary artery. Management of CAF and operative indication are still unclear. Although surgical correction is indicated when the shunt flow is significant, post operated complications such as arrhythmia, myocardial infarction due to thrombosis and cardiomyopathy were reported.
Methods and Results: We retrospectively reviewed 20 patients identified with CAF in our hospital between 2004 and 2018. Median

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

follow up duration was 10.1 ± 9.2 years. We classified Patients with CAF as proximal type 6 or distal type 14. 17 patients underwent surgical repair. Intracardiac ligation performed in 6, extracardiac ligation in 4, combined intra and extracardiac ligation in 4, transcoronary closure accompanied by coronary artery bypass graft in 2 and transcatheter closure in 1. 5 patients (distal type 1 and proximal type 4) had post operated thrombosis. 7patients of cases with post operated residual communication or conservative cases had no thrombotic events. 5 patients had mild to moderate aortic valve regurgitation (AR) including post operated cases.
Conclusions: Post operated thrombotic episodes were frequent. We must choose the operative indication and options to the type of CAFs, coronary artery dominancy and distal branchs. And Proxymal type needs attention to pouch formation and distal type to slow flow of coronary ciruculation of residual aneurysm and coronary dilatation. And anticoagulation is needed for such high risk cases. Residual small shunt may raise the possiibilty to reduce the risk of post operated thrombosis. And the careful evaluation of AR should be needed even after operation. in distal type or coronary dilatation.

P-235

Outcome after surgery for pulmonary atresia and VSD; a long-term follow study in a single institution in Sweden

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Objective: To examine the outcome after treatment of pulmonary atresia and VSD.
Methods: All children in our referral area (50% coverage of the Sweden population) operated upon due to pulmonary atresia and VSD from Jan 1st 1994 to Dec 31st 2017 were included. Patient files were analyzed and cross-checked against the Swedish National Population Registry as of Jan 1st 2018 allowing for reliable and complete data on survival. Primary outcome was all cause mortality, secondary outcomes were incidence of extracardiac diseases, reoperations and catheter interventions.
Results: Seventy patients were identified (31 girls, 39 boys) with a median age of one day (0-480) at presentation. All medical files were retrieved with no patient lost to follow-up. The pulmonary arteries were confluent in 61 (87.1%) patients, in whom a ductal supply was diagnosed in 44 (62.8%) and no ductal supply was found in 17 (24.2%). The pulmonary arteries were non-confluent in five patients (7.1%), and in four (5.7%) no central arteries were found. A right aortic arch was seen in 14 patients (20%), 22q11 deletion syndrome in 16 (22.8%) and other significant syndromes in a further nine patients (12.8%). Major aorto-pulmonary collateral arteries (MAPCAs) were found in 29 patients (41.4%), in whom 12 a unifocalization procedure was performed. Corrective surgery was accomplished in 58 patients (82.8%) at a median age of 1.3 years (0.01-19). Eight patients received a Melody® valve. Death occurred in 18 patients (25.7%), at a median age of 1.3 years (0.02-19), of whom three died within and 15 later than 30 days after the last major surgery. Mortality was similar in patients with and without MAPCAs (27.6% vs 24.4%). All deaths were cardiac except one, where the child died in an accident. The median follow-up time in survivors was 13 years (0.8-39.4) after birth.
Conclusion: Corrective surgery was accomplished in 82.8% of all patients, and long-term survival was 74.3%, with no difference found in patients with or without MAPCAs. The incidence of associated syndromes was high, with 22q11 syndrome in 22.8% and other significant syndromes in another 12.8%.

P-236

Outcome after surgical repair of atrioventricular canal defects in patients with trisomy 21

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Introduction: Trisomy 21 is often associated with heart defects and especially of the atrioventricular canal (AV-canal). The aim of the study was the analysis of outcomes after correction of AV-canal defects comparing surgical techniques, complications, and results in patients with and without trisomy 21.
Methods: We performed a retrospective study of 241 patients undergoing surgical repair of AV-canal defects during the years 1986 and 2016. One-hundred twenty-four patients (51%) had had trisomy 21 (group D). The data of these patients were compared to the data of patients without trisomy 21 (group non-D; n=117). Mean follow-up was 113 months.
Results: Twenty-one patients had undergone previous cardiac surgery; there was no significant difference between the groups (10.5% group D vs. 6.8% non-D). Trisomy 21 patients had had more often complete AV-canal defect (77.4% vs. 30% group non-D, $p<0.05$) and more often pulmonary hypertension (78% of patients in group D vs. 29% in non-D group, $p<0.05$). Down-patients were significantly younger at the time of surgery (24.8 ±56.6 months vs. 51.5±109.1 months in non-D-group) and subsequently smaller. After surgery, Down-patients required a longer period of mechanical ventilation (4.9±8.3 days vs. 1.8±3.2 days, $p<0.001$) and they needed catecholamine therapy for a longer time (2.9±3.5 days vs. 1.5±5.1 days, $p<0.05$). The 30-days mortality was comparable between the two groups (n=9 in group D vs. n=5 in group non-D, p=0.37). There was also no difference in the need for re-operation or intervention during follow-up (16.1% group D vs. 12% group non-D, p=0.35). The most common indications for re-operation were mitral valve regurgitation and left ventricular outflow tract obstruction. Echocardiographic examination at last follow-up appointment demonstrated a better mitral valve function in patients with trisomy 21.
Conclusions: Outcome after repair of AV-canal defects is very good with low re-operation rate during follow-up. Although patients with trisomy 21 had a more complex defect morphology and were younger at surgery, they have an equal chance for good outcome after surgical correction.

P-237

Therapeutic effect of medications for pulmonary hypertension in congenital heart disease after palliative surgery

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Introduction: The effect of medication for pulmonary hypertension associated with congenital heart disease is widely known. However, in clinical practice, even if it does not correspond to the definition of pulmonary hypertension (mean pulmonary artery pressure ≥ 25 mmHg), it is used for better intracardiac repair in patients after palliative surgery.
Objectives: To examine the efficacy of treatment for pulmonary hypertension in congenital heart disease after palliative surgery.
Method: The patients with palliative surgery who introduced pulmonary hypertension medication were enrolled in this study. We performed cardiac catheterization before and after introduction.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

We measured pressure gradient (ΔP) at mean pulmonary artery and left atrial pressure, pulmonary vascular resistance (Rp), PA index, pulmonary blood flow ratio (Qp / Qs), aortic oxygen saturation (SaO 2) and compared before and after. We divided the patients into two groups with mean pulmonary artery pressure ≥ 25 mmg (PH group) and <25 mm Hg (no PH group) and compared each parameters.
Result: There are 20 subjects. Major cardiac lesions are tetralogy of Fallot, atrial septal defect and ventricular septal defect. Blalock-Taussig shunt was performed in 10 cases, 6 cases of pulmonary artery banding and 4 of other surgery. Single or two types of pulmonary hypertension medication were administered and there were no cases of side effects. Rp and ΔP were significantly decreased, respectively (4.4 (1.9-15.7) to 2.4 (0.8 - 8.4) unit · m 2, 16 (6-58) to 9 (3-31) mmHg, p<0.01). PA index, Qp / QS, and SaO 2 did not change. In PH group (n = 10), Rp and ΔP decreased, respectively (5.6 (3.0-15.7) to 2.8 (0.8-5.8) unit · m 2 (p <0.05), 28 (15-58) to 13(3-31) mmHg). In no PH group (n = 10), there were no significant changes in Rp(2.9 (1.9 - 6.4) to 2.1 (1.1 - 8.4) unit · m 2), ΔP (8 (6 -16) to 8 (3 - 16) mmHg), SaO 2 and Qp / QS.
Conclusion: The pulmonary hypertension treatment after palliative surgery reduces Rp and ΔP, and may contribute to a better and safe cardiac procedure. These effects is limited to cases corresponding to pulmonary hypertension.

P-239

Vascular rings- Diagnosis and management of 32 children: Early diagnosis better outcome

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Introduction: Our aim was to investigate the presentation, diagnosis and outcome of the children with vascular rings. Methods: In this retrospective study, we reviewed the medical records of the children with vascular rings who were operated between January 2016 and June 2018. Demographic data, presenting symptoms, accompanying lesions, diagnostic methods, type of vascular ring, postoperative complications and outcome data were analyzed. Results: Mean age was 1.72 years (range 0.1 to 11), mean weight was 10.2 kg (range 2.5 to 40 kg). Seven patients (21.8%) had accompanying congenital heart defect. The most common presenting symptom was feeding difficulty (18 patient, 44%). All cases were evaluated by echocardiography and demonstrated vascular rings in 5 cases (15%). Barium esophagography was performed in 7 patients (%21.8). All patients had 64-slice CT angiography. In our centre, we are generally using 64-slice CT angiography as an imaging modality (Figure 1). CT angiography defines the type of arcus anomaly in a majority of patients and shows the anatomical relation of the arch to oesophagus and trachea. For patients who had congenital heart defects, pulmonary banding in one patient, coarctation of the aorta surgery in 5 patients, atrial septal defect surgical closure in one patient and PDA ligation in 3 patients were performed during the vascular ring surgery. Postoperative complications included: Atelectasis (three cases), chylothorax (one case), pneumonia (one case). There was no mortality. Mean follow-up duration was 2.3 years (range 0.5 to 3 years). Of these 32 patients, 23 patient (71%) was symptom-free after the first 6 months, five patients required >6 months to 1 year to reach full recovery. Four patients (12%) were still experiencing respiratory problems due to tracheomalacia. Clinical characteristics and types of the vascular ring were summarized in Table 1. Conclusions: In our series, gastrointestinal and respiratory symptoms are common. These symptoms are nonspecific in infants. Therefore, physicians should always consider the possibility of the vascular ring in children in this setting. Early diagnosis may prevent long-term respiratory issues after surgical treatment. CT angiography is an effective imaging modality to define vascular ring anomaly.

Table 1 Type of vascular rings and presenting symptoms data

Symptoms	
Respiratory (Stridor/Wheezing)	12
Feeding difficulties	18
Respiratory problems/feeding difficulties	8
Reflex apnea	1
Anatomic type	
Double arcus aorta	6
Aberrant right subclavian artery (ARSA)	8
Aberrant left subclavian artery (ALSA)	3
Pulmonary sling	2
Innominate artery compression	9
Kommerell diverticulum (isolated/combined)	5
Genetic anomalies (Down syndrome, CHARGE)	3
Associated congenital heart defects (Ventricular septal defect, Coarctation of the aorta, Patent ductus arteriosus, atrial septal defect)	7

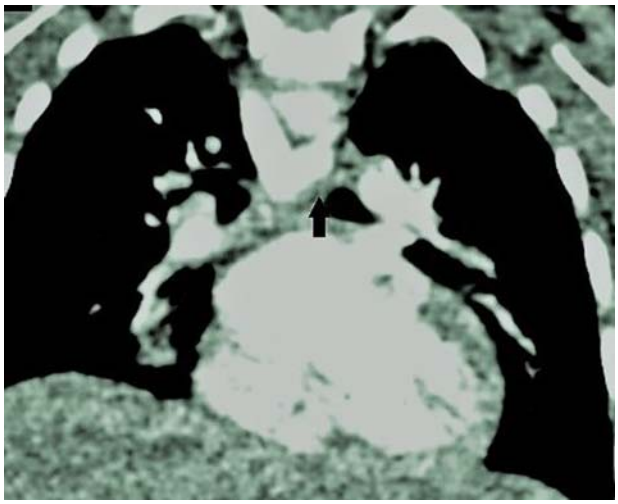

Figure 1.
ALSA + Kommerell div

P-240

Outcome after the Norwood operation for Hypoplastic left heart syndrome and related malformations – a 22 years single centre experience

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Introduction: Survival for Hypoplastic left heart syndrome (HLHS) and related single-ventricle lesions improved over the past decades. However, the Norwood circulation with its vulnerable balance between pulmonary and systemic blood flow is still associated with relatively high mortality. We aimed to identify risk factors for adverse outcome after the Norwood procedure and subsequent superior cavopulmonary anastomosis (SCPA).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Methods: Medical records of 379 patients (HLHS, n=304; non-HLHS, n=75) who underwent a Norwood procedure between 1996 and 2018 were reviewed. Follow-up to SCPA was completed in all survivors. Important changes in management included the utilization of antegrade cerebral perfusion (January 2000) and the introduction of an interstage surveillance program (October 2005). Three groups were analysed (Group 1: 1996-1999, n=50; Group 2: 2000-2005, n=103; Group 3: 2006-2018, n=226).
Results: Median age at surgery was 6 (IQR 4-9) days. A modified BT-Shunt was the standard source of pulmonary blood flow (n=357, 94.2%); 3 mm or 3.5 mm shunts were most commonly used. Early mortality (within the first 30 postoperative days or before discharge) was 10.6% and declined over time (Group 1: 22.0%; Group 2: 12.6%; Group 3: 7.1%, p=0.008). With introduction of the home-monitoring program interstage mortality of discharged patients decreased (12/117 vs. 5/147, p=0.041), but inpatient treatment before SCPA became more common (10/130 vs. 63/209, p<0.001).
Survival to SCPA was 83.2% and improved over time (Group 1: 70.0%; Group 2: 79.2%; Group 3: 90.7%, p<0.001). Weight at surgery <3 kg (p=0.041), ascending aorta <2 mm (p=0.006) and use of a 3 mm shunt (p=0.017) were identified as risk factors for mortality prior to SCPA. The indexed shunt diameter was not different between survivors and non-survivors (1.05 ±0.13 mm/kg vs. 1.08 ±0.14 mm/kg, p=0.123). Early mortality after SCPA was 6.4%. Age at surgery <90 days (p=0.014) and inpatient treatment before SCPA (p<0.001) were related to increased mortality.
Conclusions: The postoperative course after the Norwood operation and the interstage period before SCPA still carry a relatively high risk of mortality. The introduction of home-monitoring programs reduces interstage death, but children not fulfilling discharge criteria have a poorer prognosis in terms of survival after SCPA.

P-241
Hypophosphatemia following staged surgical palliation of Hypoplastic Left Heart Syndrome

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Introduction: Hypophosphatemia is commonly seen in critically ill children and has been shown to hamper clinical recovery. Patients after surgical palliation of HLHS are prone to develop this disturbance as they require large doses of medications known to decrease serum phosphorus levels. Moreover, deleterious effects of hypophosphatemia on the cardiopulmonary system can be especially harmful to those patients.
Methods: We conducted a retrospective review of the medical records of children consecutively admitted to our PICU between March 2014 to September 2018, immediately after Norwood, Glenn or Fontan procedure. The following data were recorded: age, weight, presence of malnutrition, type of procedure with assigned Aristotle Basic Complexity Score, duration of cardiopulmonary bypass, serum phosphorus and magnesium levels monitored during the first 3 days of PICU admission, hemodynamic parameters, medications, use of blood products, duration of mechanical ventilation and PICU length of stay.
Results: 89 children were included in the study, with a median age of 6,4 months (range: 2d - 75.7m). Throughout the study period decreased serum phosphorus levels occurred in 39 patients (44%), and we observed 6 cases of refrectory hypoophosphatemia, which did not respond to single potassium phosphate infusion. The mean age and weight at the time of the procedure was significantly lower for the hypophosphatemic group and the mean Aristotle Basic Complexity Score (perioperative morbidity, mortality, and technical difficulty of the procedure) was significantly higher. Whats more, epinephrine and dopamine use showed independent association with hypophosphatemia.
Conclusions: Hypophosphatemia is highly prevalent in children after staged surgical palliation of HLHS. Given the greater susceptibility and potential complications, serum phosphorus levels should be routinely measured after the surgery, so that appropriate replacement therapy may be started.

P-242
Long-term outcomes after Ross procedure in different age groups: a single - institution experience

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Objective: To evaluate long-term outcomes after Ross procedure.
Patients and Methods: All consecutive patients (n=231), operated between 1996 and 2017 at our institution, were analyzed. Among them, neonates and infants were 16 (6,9%). The mean age was 145±101 (2-648) months, mean weight - 40±22,9 (3.3-119)kg. Aortic stenosis - 111 (48%) patients, aortic insufficiency - 95 (42%) and combined lesion - 25 (10%). Aortic root reinforcement was used in 123 (53%) patients, Ross-Konno procedure in 25 (10,8%). Right ventricle-pulmonary artery (RV-PA) connection was created by prosthetic trileaflets conduits in 125 (54%), homografts – 11 (5%), xenografts – 32 (14%), RV-PA autologous tissue - 63 (27%). Long-term follow-up was 132±67 (6-252) months. Regression and correlation analysis was used to assess the results.
Results: Long-term outcomes followed in 204 (95%) patients. Overall mortality was 9,5% (n=22), hospital mortality was 6,9% (n=16) and late – 2,6% (n=6). During the follow-up, reoperations required 11 (5,1%) patients on the pulmonary autograft (4 repairs and 7 replacements), 35 (16,2%) on the RV-PA conduit (4 plasty and 31 replacement). Transcatheter procedures were performed in 40 (18,6%) patients. Independent predictors of autograft reoperation were older age (r = 0,01, p = 0,033) and preoperative aortic regurgitation (r = 0,1, p = 0,04). Independent predictors of autograft regurgitation were older age of patients at the time of surgery (r = 0,277, p = 0,003), acquired AV lesion (rheumatic disease (r = 0,252, p = 0.001), infective endocarditis (r = 0,314, p = 0,006), EDI LV>100ml/m2 before surgery (r = 0,477, p = 0.001). Factors which decrease risks of autograft disfunctions and reoperations are aortic root reinforcement (r = -0,287, p = 0.002), congenital aortic valve lesion (r =-0,405, p = 0.001), bicuspid aortic valve (r = -0,351, p = 0,001). Independent predictors of RV-PA reoperation were younger age (r = -0,174, p = 0,03) and small conduit size (r = -0,38,p = 0,001).
Conclusions: Ross procedure is an operation with low mortality and good long-term follow-up results. Reoperations on pulmonary autograft were rare in our patients. RV-PA conduit require more reoperations and it is important to continue further study.

P-243
Assessment of the pulmonary artery banding in patients with congenitally corrected transposition of the great arteries

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Objective: To assess by echocardiography the effectiveness of pulmonary artery banding in patients with congenitally corrected transposition of the great arteries (CCTGA).
Patients and Methods: From 2003 to 2017 93 patients with CCTGA from neonatal period to 68 years old were observed. Pulmonary artery banding (PAB) was performed in 22 patients with mean age of 2,72±24,56 (0,1-84) months, and mean weight of 8,5± 5,1 (3,2-22) kg.
Patients were assessed by intraoperative transesophageal echocardiography (ITEE) and transthoracic echocardiography (TEE) after the operation.
Results: Indications for PAB were: unrestricted ventricular septal defect in 14 patients (63,6%); severe tricuspid valve insufficiency with intact ventricular septum in 3 patients (13,6%); training of the left ventricle with intact ventricular septum or restrictive ventricular septal defect in 5 (22,7%) patients.
Intraoperative left ventricular pressure consisted in average of 52,3±13,73% (35%-80%) from systemic pressure. Peak pressure gradient across the pulmonary artery measured by the TTE was in average 47,9±15,9 (20-70) mmHg in early postoperative period. The next operation stage was performed in 10 (45,5%) patients. The double switch operation was performed in 8 (36,4%) patients in average of 70±53,2 months (9-144) after PAB. Peak pressure gradient across the pulmonary artery measured by the TTE was in average of 70,1 ± 13,9 (50- 92) mmHg. The bidirectional cavopulmonary anastomosis was performed in 2 patients (9%) in 10 months in the first patient and 54 months in the second patient. Peak pressure gradient across the pulmonary artery in patients with bidirectional cavopulmonary anastomosis was in average of 79,5 ± 7 mmHg by TEE data.
Conclusions: PAB is commended as a stage operation for patients with CCTGA with wide spectrum of indications. Transthoracic echocardiography is a primary method for the assessment of the pulmonary artery banding effectiveness and method for defining indications to the next stages of operation in patients with CCTGA.

P-244
Outcome of neonates with congenital heart disease born either at less than 35/40 gestation or weighing less than 2.5kg

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Introduction: Congenital heart disease (CHD) effects 8 in every 1000 babies born. Some of these neonates are born prematurely and therefore with low birth weights. In order to have the necessary intervention these neonates often have to wait until they reach an appropriate weight, generally thought of as 2.5kg. This can mean a prolonged hospital admission. The aim of this study is to gather information to help in counselling families and to identify if there are any prognostic factors that can be recognised soon after delivery.
Methods: Retrospective database analysis using the neonatal database Badgernet and the cardiac database in Leeds General Infirmary to collect data on neonates born in or treated at two tertiary neonatal units in Yorkshire between 01/01/2014 and 31/12/2016. Neonates included were born at less than 35 weeks gestation or weighing less than 2.5kg. The only exclusion was a diagnosis of patent ductus arteriosus only, neonates were included irrespective of genetic anomalies, non-cardiac comorbidity and whether or not they received intervention.
Results: 190 neonates were identified with a diagnosis of CHD and 3921 with no CHD. Neonates with CHD weighing more than 1kg are significantly more likely to die than those without CHD (29.5% v's 1.6%, $p<0.01$). There was no significant increase in mortality in the CHD group in those with a chromosomal abnormality than in those without (37.5% v's 27%, $p=0.22$) however those that died carried a huge burden of co-morbidity with 95% displaying at least 1 other significant non-cardiac system disorder (median 2, range 0-4). Neonates with CHD were significantly more likely to develop necrotising enterocolitis than those without CHD (12.6 v's 6.9%, $p<0.01$). When looking at length of stay 56% of neonates with CHD were discharged within 50 days of birth, 18% were admitted for over 100 days and 3% for over 200 days.
Conclusions: This study enables parents to have a more realistic idea of what their babies neonatal course will be from length of stay to mortality and morbidity during admission. This can be invaluable at such a stressful time.

P-245
Hybrid melody valve implantation in mitral position in a child with Shone's complex and failure of two previous prosthetic valves

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Background: The use of the stented bovine jugular vein graft (MelodyTM valve) in the mitral position in children is an off-label treatment option for valve replacement. It is useful as not only it can be implanted in very small annuli but also it may limit pannus formation. Additionally only antiplatelet treatment is sufficient postoperatively, compared to anticoagulation for mechanical valves.
Case Report: We describe a case of MelodyTM valve implantation in the mitral position in a three year old girl with Shone's complex who underwent two unsuccessful mitral valve repairs followed by two mechanical valve replacements. Both mechanical valves prosthesis were 19 mm Saint Jude®, placed supraannular. During removal of both mechanical valves, pannus formation was noted intraoperatively, due to florid fibrous proliferation presumably related to underlying Shone's complex. This pannus was impinging the valve mechanism with restricted leaflet motion on fluoroscopy and there was secondary thrombus formation.
It was felt that further mechanical valve replacement would lead to the same problem. An 18 mm MelodyTM valve was chosen and implanted surgically as per the technique described by the Boston Group (Shortening and trimming of the valve, creating a wide V shape opening to the outflow). The MelodyTM valve was then directly balloon expanded up to 18 mm. The heart resumed activity in complete heart block needing pacing. Later a permanent pacemaker was implanted. The immediate post-operative Echo showed valve length 23 mm with circumferential inner diameter 16mm. The mean estimated inflow gradient through the MelodyTM valve was 4 mmHg with no regurgitation. The peak velocity in the left ventricle outflow tract was 1.6 m/sec. The postoperative period was uncomplicated and the child was discharged after 20 days.
Conclusion: In selected patients MelodyTM valve should be considered as a treatment option for mitral valve disease in children especially where all other techniques have failed. This technique as described by the Boston Group can be safely performed even in complex cases such as this one. This use of the MelodyTM valve, is however off-label without long-term follow-up data, and so close follow-up of the patient is needed.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Outcome after primary repair of Tetralogy of Fallot (TOF) – A single center retrospective analysis of long-term results in 176 patients

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Objectives: The aim of our study was to analyze predictors for early and late mortality, need for re-intervention or re-surgery after TOF repair depending on the surgical strategy for primary repair (rTOF).
Methods: All patients, who underwent rTOF in our center between April 1999 and January 2016 (n=176) were included. 104 male and 72 female patients were assessed (0-4 months, n=51; 4-8 months, n=77; 8-12 months, n=24; >1 year, n=24). Mean body weight was 6.6 (2.5-70) in males and 6.4 (2.9-56) kilograms in females. The surgical strategy was transannular patch (TAP) (n=80), valve-sparing double-patch technique (BiPatch) (n=92) or graft implantation (n=4), The Surgical approach was either transatrial (n=10) or transventricular (n=166). Patients with initial palliation by shunt were excluded. The protocol was approved by our ethics committee.
Results: The need for re-surgery was not influenced by the surgical technique. During the observation period of 120 months 81.3% of the cohort did not need re-surgery (77.5% in TAP-group, 75% after graft implantation and 84.8% after BiPatch repair). Overall survival after rTOF was 97.7%. The need of re-surgery was not influenced by the complexity of the patients (Aristotle Complexity Score). We also found no significant influence of neither age nor sex, body weight or height. Indications for surgical re-intervention were more than moderate pulmonary regurgitation (TAP n=45, BiPatch n=26) or stenosis (TAP n=3, BiPatch n=136). In terms of the neurological outcome there was no significant difference between patients who had underwent TAP or bi-patch repair.
Conclusions: In our cohort we found no significant difference in respect to the outcome and need of re-intervention in TOF patients depending on surgical technique, age, body weight, gender or complexity.

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Before and after: Interventions and Tetralogy of Fallot repair

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Introduction: Risk factors for re-intervention after repair of Tetralogy of Fallot (TOF) include primary repair at less than 28 days of age, small pulmonary arteries and a staged operative strategy. Recent evidence has suggested that palliation to the right ventricular outflow tract (RVOT) is associated with increased re-interventions. We aimed to determine whether having a catheter or surgical procedure prior to repair resulted in an increased risk of post-repair interventions in our patient population.
Methods: Retrospective single centre study. Inclusion criteria were all patients who had undergone TOF repair between 1/1/2007 to 31/12/2017. Data was obtained from the hospital database. Exclusion criteria were those with pulmonary atresia and ventricular septal defect variants. Patients were divided into a primary repair group and a staged group. Prior intervention was specified as either catheter intervention or surgical shunt. Re-interventions post repair included redo surgery, further catheter interventions or pulmonary valve replacement.
Results: In the study period 219 patients underwent TOF repair. 179 (82%) patients had primary repair at a mean of 252 days of age (range 6-2844). 39 patients (18%) were in the staged group and had 1 or more interventions prior to full repair, occurring at a mean of 321.47 days (range 53-1523). In the staged group 15(38%) had systemic to pulmonary artery shunt, 24(62%) had a catheter intervention. Mortality following full repair was 0% at 30 days. There were 3 late mortalities at 4, 5 and 17mths. Re-intervention following full repair was 38 (22%) in the primary repair group and 1 (45%) in a staged group ($p<0.002$). Neither placement of a transannular patch ($p=0.21$) or having a previous shunt ($p=0.17$) were associated with increased risk of interventions post repair. Pulmonary valve replacement was required in 7 patients in the study period, all of whom had undergone primary repair.
Conclusions: Patients who underwent a staged approach to TOF repair were more likely to require further intervention post repair. This was especially evident in those undergoing a catheter intervention. This is important information when planning treatment and for counselling families regarding future interventions and their timing.

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Early life Predictors for Major Adverse Events in Hypoplastic Left Heart Syndrome after Norwood Stage I Palliation: A 25-year Retrospective Study

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Objective: We aimed to identify perinatal risk factors for severe complications in infants with hypoplastic left heart syndrome (HLHS).
Methods: Ninety-three patients with HLHS who underwent stage I Norwood palliation between 1993-2017 were included. Fetal and postnatal echocardiograms, echo notes, demographic, surgical and other clinical data were reviewed. Fetal and postnatal risk variables included restrictive or intact atrial septum (RAS/IAS), left deviation of septum primum (LDSP), significant tricuspid regurgitation (sTR), right ventricle (RV) dysfunction, left ventricular (LV) morphology (aortic stenosis (AS)&mitral stenosis (MS), aortic atresia (AA)&MS and AA&MA), postnatal diagnosis and low preoperative weight (LPW; ≤2.5 kg). Major outcome variables were survival, need for ventricular assist device (VAD) and ECMO, protein-loosing enteropathy and heart transplant (Htx).
Results: The overall survival was 67% (interstage I: 76%; interstage II: 91%; post-TCPC 95%), rising from 54% during 1993-2002 to 81% during 2003-2017. Fetal diagnosis was available in 47%, rising from 22% during 1993-2002 to 64% during 2003-2017. Intrauterine sTR was linked to higher pulmonary pressure at stage II (15.7±2.3 vs. 11.8±2.8; $p=0.03$) and lower interstage II survival (67% vs. 96%, $p=0.06$). Postnatal diagnosis was associated with initial RV dysfunction ($p=0.007$), sTR ($p=0.005$) and delayed stage I surgery ($p=0.001$) but did not influence survival. LDSP was diagnosed in 71% of infants with RAS and was most often linked to AA&MA ($p=0.0004$), whereas AA&MS was associated with lower interstage I (60% vs. 84%; $p=0.01$) and overall survival (53% vs. 73%; $p=0.06$). RAS/IAS was linked to atrial septostomy prior

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

to stage I (p<0.0001), longer mechanical ventilation (p=0.04), ICU (p=0.002) and overall stay in hospital (p=0.0002) after stage I as well as lower survival after stage III (p=0.006). LPW at stage I was associated with lower interstage I survival (45% vs. 80%; p=0.01), need of VAD/ECMO (2/11 vs. 2/82; p=0.02) and Htx (2/11 vs. 0/82; p<0.001).
Conclusion: In patients with HLHS, certain LV anatomical subtypes along with restrictive atrial septum and low body weight at stage I remain important risk factors for severe complications later in life.

P-250
Surgery over stents. New challenges to face

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Introduction: 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: 111 stents in 90 patients were partial or totally removed at surgery. Univentricular heart was diagnosed in 40 patients. Stents were previously deployed in: ductus (17), right ventricle outflow tract (RVOT, 28), atrial septal defect (ASD, 10), right pulmonary artery (RPA, 13), left pulmonary artery (LPA, 29), inferior vena cava (IVC, 6), superior vena cava (SVC, 5) ascending aorta (AAo, 2) and pulmonary veins (1). Surgical procedures performed: 19 transplants, 11 Fontan, 6 Glenn, 2 comprehensive repair (Norwood + Glenn), 2 Glenn take-down, 17 conduit replacement, 11 Fallot, 7 Rastelli, 2 Ross-Konno, and others (11).
Results: Ten ductal stents were clipped. 29 stents in RVOT, 9 in ASD, 7 in RPA, 14 in the LPA, 7 in IVC, 2 in SVC one in the ascending aorta and one in the pulmonary veins were completely removed. Seven stents in RPA, three in SVC, one in AAo and seventeen in the LPA and two in the RVOT 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.

P-251
Customized approach in minimally-invasive pediatric cardiac surgery

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Introduction: A minimally-invasive pediatric cardiac surgery set-up program is described. Children are offered a customized approach according to cardiac condition plus weight. Data are collected in a prospective way, spanning from January 2013 until November 2018.
Methods: Simple cases are scheduled to begin with (atrial and ventricular septal defects). Surgical approach was body-size dependent: lower mini-sternotomy for children under 10 Kg. (ventricular septal defects, mainly), postero-lateral thoracotomy in kids between 10-25 Kg. and sub-mammary crease incision over 25 Kg. (teens and young adults). On gathering experience, more cardiopathies were gradually introduced. The postero-lateral approach was shifted to an axillary one and the sub-mammary incision was swapped to a video-assisted mini-thoracotomy (with peripheral cannulation). The upper mini-sternotomy was added to fix aortic valvulopathies.

Table 1: surgical minimally-invasive approaches

APPROACH	NUMBER
Sub-mammary	35
Axillary	43
Lateral-posterior	11
Upper mini-sternotomy	22
Lower mini-sternotomy	150
Video-assisted mini-thoracotomy	20
TOTAL	281

Table 2: cardiac conditions approached

CARDIAC CONDITIONS	NUMBER
Ostium Secundum Atrial Septal Defect (ASD)	100
Sinus Venosus ASD	23
Ventricular Septal Defect (VSD)	84
Ostium Primum ASD (Partial AV septal defect)	22
Complete atrio-ventricular septal defect	8
Aortic valve stenosis/regurgitation	21
Others	23
TOTAL	281

Results: 281 patients were operated on: 100 ostium secundum atrial septal defect, 23 sinus venosus atrial septal defect, 84 ventricular septal defect, 22 partial atrio-ventricular septal defect, 8 complete atrio-ventricular septal defect, 21 aortic valve repair and 23 miscellaneous. Surgical approaches selected were: 150 lower-mini-sternotomy, 35 sub-mammary, 11 postero-lateral thoracotomy, 43 axillary, 22 upper mini-sternotomy and 20 video-assisted mini-thoracotomy. The ratio of minimally invasive surgery to full sternotomy increased from 20% in 2013 to 30% in 2018.
Conclusions: A customized approach (cardiopathy and weight differentiated) in a new program for minimally-invasive congenital heart surgery proved successful. Moving to smaller incisions and adding new diagnosis is feasible as long as the surgeons become more proficient with the technique. Consequently, the percentage of less invasive approaches raises in a short lapse of time.

P-252
Postcardiotomy ECMO in pediatric patients: excellent outcomes in a contemporary series

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Introduction: A percentage of patients undergoing cardiac surgery requires a circulatory and respiratory support with an Extracorporeal Membrane Oxygenation (ECMO) device for a variable period, due to an insufficient reaction to maximal conventional therapy in the immediate postoperative period.
Methods: From 2009 to 2018, 80 pediatric patients required ECMO in our institution. 44% of them were new-borns, with an average age of 15.4 months and a weight of 6.9 Kg. The most common univentricular physiology was HLHS (6 patients), whereas regarding biventricular correction it was Transposition/Taussig Bing Type (9), followed by biventricular correction of

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Hypoplastic Left Heart Complex (8). The main indication (60%) was an impossibility to wean the patient from Extracorporeal Circulation.
The variables that have been analyzed were: sex and age of the patients, cardioplegia solutions types, neonatal age, patients physiology, extracorporeal circulation and cross-clamp time, left vent drainage, hemofiltration in the postoperative period, reinterventions during ECMO assistance, long-term ECMO (> 5 days), Ecmo indications, need for a second ECMO after weaning from the first one, postoperative complications.
Results: Of the 80 patients, weaning from the ECMO was achieved in 78% of the patients, with a survival rate of 66%. The Last ELSO data of July 2018 report 68% of weaning with a survival rate of 52%.
The risk factors associated with mortality in the univariate analysis were: Neonatal age (p=0,013), univentricular physiology (p= 0,004), hemofiltration in the postoperative period (p=0,011) and between the postoperative complications, only the neurological ones (p=0,05).
On the multivariate analysis, risk factors were Neonatal age (p=0,02) and Univentricular Physiology (p=0,03).
During the Ecmo period, there were noticed residual lesions that needed some kind of interventions (by surgery or catheterism) in 47% of the patients.
Conclusions: We herein report a series of postcardiotomy ECMO in pediatric patients with good results in terms of weaning and survival. Almost half of the patients needed some kind of reintervention, hence it is very important looking for postoperative residual lesions.

P-253
Aortic valve surgical repair in congenital patients: does age and size really matter?

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Objectives: Congenital aortic valvulopathy is a real challenge for the valve sparing surgery. These valves compared with acquired pathology are very complex, due to structural anomalies of the valve itself and the associated lesions at the sub/supravalvular levels or the aortic root. The patient´s size (from infant to adult) is also a problem. In order to delay the time of the valve replacement, our first surgical option in each patient is to spare his aortic native valve. We report the conservative aortic valve surgery in our congenital heart unit and analyse the differences between children and adults patients.
Methods: Retrospective study: 61 operations performed in 59 patients, during the period 2010-2018. Two groups of 46 children (<14 years) and 15 adults (≥14 years) were compared. Statistical analysis was done with SPSS-15.0.
Results: Median age in children-group: 1,5 years, in adults-group 28 years. Previous surgery was more frequent in adults-47% compared with children-26% (pns). Children have mainly a stenotic aortic valve (bicuspid-46%, monocuspid-15%), and adults have a dilated aortic root with preserved valve function (p: 0,04).
Operations performed with extracorporeal circulation, aortic clamp and trans-aortic approach. Surgical techniques were different in each group (p:0,007): Children valvuloplasty included comissurotomy-6(13%), aortic leaflet plasty-16(35%), comissurotomy + aortic leaflet plasty-23(50%), and David operation-1(2%). Adults received David operation-6(40%), aortic leaflet plasty-5(33%), and comissurotomy + aortic leaflet plasty-4(27%). Associated surgery was done in 80%-adults and 67%-children (pns). Median extracorporeal and aortic clamp times were longer in adults respect to children (p:0,02).
One child with several complications died. Inhospital mortality was 2,2% in children and 0% in adults. We had no late mortality after discharge.
Follow-up was complete, with median of 22 months (IQR: 13-39). Percutaneous reintervention was needed in 5(11%)-children and 1(7%)-adult (pns). Reoperation was required in 6(14%)-children and 1(7%)-adult (pns). Nowadays the majority of our patients in both groups are asymptomatic, with normofunction of their aortic valve.
Conclusions: Aortic valve sparing surgeries in children and adults performed in our congenital cardiovascular unit have good results, with minimal mortality and acceptable reintervention rate. Children presents mainly with stenotic aortic valves and adults with a dilated aortic root.

P-254
Outcome of atrioventricular septal defects with single ventricular palliation: 50 years of experience

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Introduction: "Single ventricles" with atrioventricular septal defect (AVSD) include a variety of complex congenital heart defects such as unbalanced AVSD and univentricular hearts with common atrioventricular valve, commonly associated with heterotaxy. Our objective was to describe hard outcomes (death or heart transplantation) and their predictors in these defects.
Methods: We conducted a retrospective, monocentric review of children with common atrioventricular valve, who entered a univentricular path repair at our institution.
Results: 161 patients with AVSD were included into the study: 121 (75%) had univentricular hearts with heterotaxy syndrome, and 40 (25%) had unbalanced AVSD that were not suitable for biventricular repair. Median follow-up was 7.3 years (max 34 years). 41.6% patients had a prenatal diagnosis. Overall mortality was 50.3%. 24% (39/161) never had surgery with a mortality of 77% at a median age of 1.7 years. 24% (39/161) had the first stage palliation (Blalock-Taussig shunt 10.5%, banding of pulmonary artery 10.5%, other 3%) with a mortality of 69% at a median age of 3.2 years. 26% of patients reached the second stage palliation (Glenn surgery) (42/161) with a mortality of 48% at a median age of 8 years, and 26% of patients had the third stage palliation (Fontan completion) (41/161) with a mortality of 10% at a median age of 16.2 years. Protective factors were prenatal diagnosis (HR 0.32, p=0.002) and to have reached Fontan stage for those who effectively entered the surgical program (HR 0.08, p<0.0001). These results were confirmed by multivariate logistic regression analysis. 70% of survivors (n=51) were in NYHA functional class I and 78% received at least one cardiac medication.
Conclusion: Mortality and morbidity of univentricular hearts with atrioventricular septal defect remain high. Prenatal diagnosis is a protective factor. Once the total cavopulmonary connection had been performed, the outcome is comparable to that of other univentricular hearts.

P-255
Biventricular intracardiac thrombi in acute myocarditis: case report

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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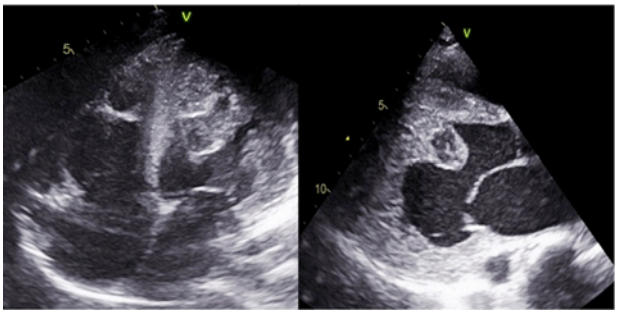
Introduction: Myocarditis is an inflammatory disease of the myocardium, presenting unusually with LV systolic dysfunction and dilatation. Although ventricular thrombus formation is rare, it carries the risk of serious complications.
Clinical Case: A previously healthy 17-year-old boy, with no relevant familiar history or medications, was admitted to the emergency department due to one-week history of acute gastroenteritis (AGE) and severe asthenia. Clinically, he presented unwell, with tachycardia and tachypnoea, normal blood pressure, tender abdomen, normal cardiac auscultation and decreased respiratory sounds. The blood samples revealed hyperlactacidemia, no leukocytosis, CRP 200mg/L, acute kidney injury, cholestatic hepatitis, elevated d-dimers as well as myocardial necrosis markers and Pro-BNP 14585pg/mL. The diagnosis of acute myocarditis in the setting of AGE was confirmed. The echocardiogram showed a severe biventricular systolic dysfunction, with moderate mitral regurgitation, several echogenic masses adhering to the walls and apex of both ventricles, suggestive of thrombi, and no signs of vegetations or pulmonary hypertension. He was admitted in the PICU and anticoagulation therapy was started. Genetic predisposition of hypercoagulability and rheumatologic disorders were ruled out. Stool sample was positive for Campylobacter jejuni. Thoracic CT excluded pulmonary thromboembolism. Despite the anticoagulation institution, a gradual and dramatic growth of the LV thrombi with LVOT extension and none cardiac function improvement were observed in the following echocardiograms. At this point, the patient presented with multiple limb arterial embolism. He was submitted to surgical thrombectomy and mitral restrictive annuloplasty with subsequent need of VA-ECMO support. Eleven days after being listed, he underwent an orthotopic heart transplant with favourable surgical and postoperative course. Endomyocardial biopsies and histologic examination diagnosed a lymphocytic myocarditis. The patient was discharged with normal LV contractility, no new thrombi formation and normal neurologic evaluation.
Conclusion: This case highlights the rarity of biventricular thrombi presentation with acute myocarditis and its unresponsiveness to anticoagulation. Surgical thrombectomy is a high-risk option in the management of these patients, with predictable drawbacks such as further ventricular dysfunction and embolism. We also present this case for its good outcome in terms of use of ECMO as a bridging therapy to heart transplantation, with no major complications.



P-256

Recurrent cardiopulmonary arrests in an infant with PA with VSD and MAPCAs: case report

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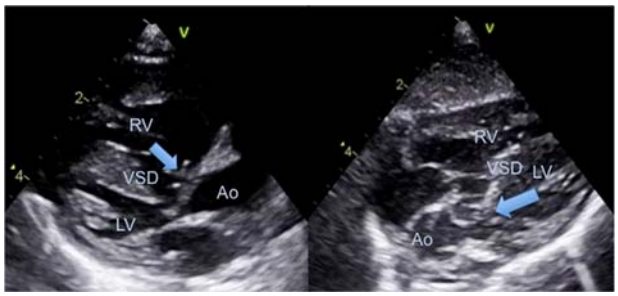
Introduction: Pulmonary atresia (PA) with ventricular septal defect (VSD) and major aorto-pulmonary collateral arteries (MAPCAs) is a rare congenital heart disease with a poor prognosis. Its management and treatment options heavily rely on the pulmonary blood flow pattern.
Clinical Case: A 2-month-old infant, postnatally diagnosed with PA/small-VSD/MAPCAs and confluent, but extremely hypoplastic Pas, was admitted to the emergency department due to dyspnoea, cyanosis (SpO2~50%) and hypotonia. She had a poor peripheral perfusion and the auscultation denoted only a continuous murmur, with the absence of the usual predominant systolic murmur. The echocardiogram, besides the known anatomy, evidenced a restrictive sub-aortic VSD, partially closed by aneurysmal tricuspid valve (TV) tissue protruding below the aortic valve, with an exclusive R-L shunt (70mmHg of gradient). A cardiac catheterization was performed, revealing multiple MAPCAs from supra-aortic branches and descending aorta to both lungs and hypoplastic PAs (nearly 1.7mm), no pressures in the LV were obtained. In the following days, she presented new episodes of hypoxemia and bradycardia while crying and subsequent cardiac arrests, requiring short courses of CPR. During the last, an echocardiogram was performed, showing the TV tissue protruding through the VSD into the left ventricle outflow tract (LVOT) limiting the flow through the aortic valve. This fact seemed to be the most reasonable explanation for the previous events, which appeared to be the cause of the recurrent cardiac arrests. She underwent a cardiac surgery with construction of a Melbourne shunt and fixation of the septal leaflet of TV to the right side of interventricular septum. The postoperative echocardiograms showed an adequate patency of the Melbourne shunt, small VSD (RV-LV gradient of 62 mmHg) and no LVOT obstruction. No more events occurred and the infant was discharged on oral aspirin with SpO2 75-80% and normal neurological evaluation. Future surgical intervention is still under discussion.
Conclusion: This case claims the attention to a rare cause of cardiac arrest in the setting of a PA with VSD, caused by dynamic LVOT obstruction.



https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-257

Don't Miss the Coronary Arteries

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Introduction: coronary arteries abnormalities is frequently associated with severe aortic stenosis. It has been hypothetised that the same ischaemic noxa that resulted in aortic valve stenosis may be the cause of coronary pathology. Surgical complications related to coronary artery abnormality result in 2% morbidity in the inmediate postoperative period.
Case Report: We present a clinical case of an 11 years old patient with severe aortic valve stenosis who underwent 3 percutaneous aortic valve valvuloplasty interventions. He was admitted for Ross-Konno surgical intervention. Postoperative transesophageal echocardiography evidenced aquinetic and dilated right ventricle, with a new residual severe tricuspid regurgitation. During Surgical inspection of the coronary arteries, the right coronary ostium was found to be small. The patient was admitted to PICU and due to low cardiac output syndrome, the patient was placed on ECMO. After 7 days the RV function was still low. The ECMO was changed for a right ventricle assistance. he was elected to undergo a right coronary bypass with the internal mammary artery, however it was not possible to perform due to epicardial fibrosis. 16 days later the RV function was only discreetly better so a new catheterism was done to treat the stenosis with angioplasty. 6 days after that the assistance was suspended and now the patient is discharged, at home, treated with oral diuretics and with ambulatory follow up.
Conclusions: Surgeries that involve the aortic valve can modify the coronary artery anatomy and therefore its perfusion, even when the coronaries remain untouched. Therefore, it is necessary to describe very well its anatomy not just before surgery, but also after it. Particularly if in the postoperative period the patient does not evolve as expected and other medical and surgical reasons are discarded. An early catheterization and a more aggressive approach may help to reduce morbility in these patients. Reports of individual cases may help in this condition.

P-258

Experience of cardiovascular surgery for congenital heart disease associated with trisomy 13 &18

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Objectives: this study was to examine a single institutional experience of patients with trisomy 13 and trisomy 18 in the setting of comorbid congenital heart disease (CHD) and present the outcomes of surgical intervention.
Methods: Patients with CHD and trisomy 13 or 18 presenting to our institution from 2009 through 2018 were retrospectively reviewed. Ten consecutive trisomy 18 patients and three consecutive trisomy 13 patients (13 patients in total) with comorbid CHD underwent surgical intervention. These 13 patients had ages ranging from 3-389 days (median 45 days), birth weight of 1378–3208 g (median 1972g), and length of gestation of 34–41 weeks (median 37). Palliative operations performed in 12 patients of ventricular septal defect (VSD), including pulmonary artery banding in 9, bilateral pulmonary artery banding for the treatment of HLHS variant in 1, and systemic-to-pulmonary shunt for the treatment of TOF/pulmonary atresia in 2 patients. Concomitant procedures included patent ductus arteriosus closure in 8 patients and a lung biopsy in 5 patients and were performed at a median age of 45 days (3-389 days) and a median weight of 2.15 kg (1.3-5.8 kg). Two patients underwent subsequent second-stage intracardiac repair electively. Primary intracardiac repair consisted of VSD closure and patent ductus arteriosus closure in one patient.
Results: The hospital mortality was 15% (n = 2), and 11 patients (85%) were discharged home with improved symptoms. The survival from surgical intervention ranged from 6-115 months (median 31 months). Two patients experienced late death due to non-cardiac events (respiratory failure and hepatoblastoma in one patient each).
Conclusions: Our data suggest that cardiac surgery may improve the survival in select patients with trisomy 13 and 18. However, the indication for surgery should be carefully considered on a case-by-case basis, as there remains a risk of late death even after surgery. Even if the patient can be discharged from the hospital after surgery, the family may often be overwhelmed by the care, such as respiratory support, tube feeding and have severe mental retardation for multiple congenital anomalies.

P-259

Impact of pulmonary artery banding on the common atrioventricular valve in complete atrioventricular septal defect

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Objective: The impact of pulmonary artery banding (PAB)-induced changes in hemodynamics on the common atrioventricular valve (CAVV) have not been elucidated. The purpose of this study was to clarify the impact of PAB on the CAVV in CAVSD.
Methods: The subjects were 24 patients who underwent PAB for CAVSD between 2000 and 2017. We examined changes in CAVV regurgitation (CAVVR) and left atrioventricular valve (LAVV) size growth from before PAB to before intracardiac repair (ICR). The LAVV diameter was measured on echocardiography and expressed as a z-score. The degree of CAVVR was evaluated as follows: none, 0; slight, 0.5°; mild, 1°; moderate: 2°; and severe, 3°.
Results: Leaflet anomaly was present in 4 patients. An analysis was performed for 23 patients with biventricular repair. The median age and body weight at PAB were 6.0 months (0.5–38 months) and 3100 g (1000–4615 g). The median band circumference was body weight (kg) + 17 mm (10–22 mm). The median pressure at PAB was 3.6 m/s (2.2–4.5 m/s). The median PAB-ICR period was 14 months (4–109 months). According to changes in CAVVR, the patients were classified into an exacerbation (n=4) or non-exacerbation group (n=19). In the non-exacerbation group, 15 patients (78%) showed improvement in CAVVR and the mean score before PAB was 1.4 ± 0.4° and that before ICR was 0.6 ± 0.4°. A group comparison showed no significant difference except for the leaflet anomaly (p=0.01) in all 4 patients in the exacerbation group, whose left atrioventricular valve diameter did not increase (mean z-score before PAB, −0.40±0.48 and before ICR, −0.47±0.22). In the non-exacerbation group, the LAVV diameter increased in 16 patients (84%; mean z-score before PAB, −0.47±0.90 and before ICR, −0.38±0.79). In the multivariate analysis of the LAVV size growth in all 23 patients, significant associations were observed only among leaflet anomaly (p=0.02) and CAVVR exacerbation before ICR (p=0.02).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusion: The patients without leaflet anomaly showed no CAVVR exacerbation due to the pressure load produced by PAB and showed improvement in CAVVR due to decreased ventricular volume load, suggesting the possibility of growth of the LAVV.

P-260
Additional flow on Glenn procedure enlarges not pulmonary artery size but ventricular volume after Fontan

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Introduction: We fundamentally prepare additional flow (ADF) to pulmonary arteries on Glenn procedure. We performed ADF in the form of narrowing down original left-to-right shunts to small size. However, ADF, which is left-to-right shunt, is suggestive of imposing over-loads against mono- ventricle. We predicted ADF on Glenn procedure would leave cardiac overloads even after Fontan procedure.
Methods: The medical records of 171 Fontan patients were reviewed who underwent periodic cardiac catheterization after Fontan between 2010 and 2017. We defined ADF patients as having confirmed ADF on pre-Fontan cardiac catheterization (n=111). We compared post-Fontan cardio-pulmonary indexes between Fontan patients with and without history of ADF.
Results: Additional flow was reserved by following methods, such as shunting from aortic branch (52), antegrade flow from ventricular outflow tract (38), and conduit from right ventricle (21). As for cardiac performances, ventricular volumes were larger in ADF group on end-systole (29 vs. 24 ml/m2: p=0.012) and on end-diastole(65 vs. 56 ml/m2: p=0.014). Pulmonary-capillary wedges pressures were also elevated in ADF group (6.7 vs. 5.8 mmHg: p=0.022). There were no significant differences in other cardiac performances. As for pulmonary circulation, there were no significant differences in following factors between two groups; aortic saturation oxygen, indexes of pulmonary arteries(232 vs. 220 mm2/m2), central venous pressures, and pulmonary resistances. Concerning internal use for heart failure, the patient rate with vasodilators was not different between two groups; that with beta blockers was also not different. Similarly, the amounts of enarapril and carvedilol were not different between two groups.
Conclusion: Our study showed ADF patients after Fontan possessed larger ventricular volumes and elevated capillary-wedge pressures than non-ADF patients. However, ADF patients could not gain larger pulmonary sizes after Fontan. This indicated left-to-right shuns (ADF) on Glenn circulation, whose flows we considered low, revealed their adverse effects even after Fontan procedure. Besides, ADF patients did not possess larger pulmonary-artery regardless of holding larger ventricular volumes. If we have to perform ADF to Glenn patients, we should administer heart failure therapies more proactively.

P-261
High pulmonary-artery pressures in Glenn circulation are not provoked by scanty pulmonary vascular-bed but by elevated afterloads due to decreasing cardiac functions

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Introduction: We commonly consider that small pulmonary artery is connected with high pressures of superior vena cava (SVCP) in Glenn patients. So we employ a strategy of pulmonary high flow to grow pulmonary arteries as much as possible. However, we predict pulmonary high flow increases loads against heart, which would become afterloads against pulmonary arteries. These high afterloads would end up increasing SVCP. We investigated related factors to high SVCP in Glenn circulation.
Methods: The medical records of 139 Glenn patients were reviewed who were aged 9 months to 10 years. They underwent cardiac catheterization between 2010 and 2017. We defined High SVCP as SVCP 16 mmHg or over (n=34). Cardio-pulmonary indexes were determined which were connected with high SVCP.
Results: Pulmonary artery indexes were not different at all between two groups. The rate of patients with high levels of NT-pro BNP (≥ 800 pg/ml) was higher in Glenn patients with high SVCP. High SVCP in Glenn patients was significantly related to 5 cardio-pulmonary factors: end-diastolic ventricular volumes ≥ 175% (p=0.011), end-diastolic ventricular pressures ≥ 12 mmHg (<0.0001), pulmonary capillary-wedge pressures (or left atrium pressures) ≥ 10 mmHg (<0.0001), pulmonary flow > 3.3 L/min/m2 (p=0.035), and existence of additional flows on Glenn procedure (p=0.010). After multivariate analysis, high SVCP was independently associated with odds ratio of 7.6 for elevated pressure of pulmonary capillary wedge/left atrium pressures, 4.6 for increased ventricular pressures on end-diastole, and 3.7 for additional flow to pulmonary arteries. Explanatory coefficient was high (0.48) by these 5 factors. The ratios of internal medicines for heart failure, such as vasodilators and beta blockers, were not different between Glenn patients with and without high SVCP; amount doses of enalapril and carvedilol were not different at all between two groups.
Conclusion: Our study showed increased afterloads against pulmonary arteries which were caused by cardiac overloads provoked High SVCP in Glenn patients. So the strategies that produced pulmonary high flow to grow pulmonary arteries can introduce High SVCP in Glenn patients. In the case when we could no lessen pulmonary flow, we should set intense medical treatment for cardiac afterloads in advance.

P-262
Redux Pulmonary Valve Replacement Surgery for Conotruncal Diseases

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Introduction: The repair of conotruncal diseases is often the cause of ulterior pulmonary valvular lesions, which could be life-threatening in adulthood. Iterative Pulmonary valve replacement is an obligation because the inevitable degeneration of the bioprostheses used. The goal in these patients is to delay the surgical recovery but without reaching the stage of irreversible lesions. Many questions arise regarding the benefit of this surgery and the factors on which it depends. The objective of our work was to evaluate the results of pulmonary valve replacement in the short, medium and long term.
Methods: This was a retrospective study, conducted between January 1999 and March 2016, involving 34 patients with conotruncal heart disease. They had at first a complete repair of their pathology followed in a second time of a pulmonary valve replacement.
Results: The mean age at pulmonary valve replacement was 20.22 years. The rate of patients with tetralogy of Fallot was 64.70%. The reason for valvulation was severe pulmonary regurgitation in 76.50% of cases. Pulmonary valve replacement was done by a bioprosthesis in 56% of patients and by a valved tube in the remaining patients.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Our study showed an early mortality rate of 2.9% and a rate of major postoperative complications of 32.35%. After an average follow-up of 3.14 ± 2.42 years, the results of our study were in favor of a non-significant improvement ($p = 0.36$) in the functional status of patients, the overall stability of right ventricular function ($p = 0.08$) and left ($P = 0.6$) and the decrease in QRS duration ($p = 0.05$). The survival rate without degeneration at 5 years and at 10 years was respectively 62.4% and 52%. The survival rate without second valvulation for the same durations was respectively 87.4% and 72.8%.
Conclusion: Pulmonary valve replacement is a low-risk surgery despite its iterative nature. Highly specialized management and optimization of operative timing is required in these patients, whose numbers are growing rapidly.

P-263
The Nikaidoh Procedure (Aortic Root Translocation): Early and Midterm Outcomes
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Introduction: For patients presenting with transposition of the great arteries (TGA), ventricular septal defect (VSD), and pulmonary stenosis (PS), aortic translocation or the Nikaidoh procedure has been shown to be a valuable surgical option. Despite promising early results, reports on surgical outcomes and follow-up are scarce. Herein, we report our early- and follow-up results after the Nikaidoh procedure.
Methods: Demographic, procedural, and outcome data were retrospectively collected for 11 consecutive patients who underwent the Nikaidoh procedure between 2007 and 2018 at our institution. The postoperative clinical course and need for reinterventions were reviewed, as well as echocardiograms of 9 patients.
Results: The main diagnosis was TGA/VSD/PS (n=10). Median age at operation was 7 months (range 2-36 months). The aortic root was transplanted as a free autograft followed by coronary reimplantation. Right ventricle to pulmonary artery (RV-PA) continuity was established with a valve in 5 patients (45%), and valveless in 6 patients (55%). All patients survived the operation (median ICU stay 7 days), and follow-up period (median 6.2 years, range 0.7-11.6 years). All patients show good functional capacity (NYHA class I-II). Reinterventions were performed in 5 patients. Freedom from re-intervention on the RV-PA connection was highest in patients with a valveless RV-PA connection ($p = 0.046$). No reinterventions were performed on the left ventricular (LV) outflow tract.
Echocardiography demonstrated unrestricted flow across the LV outflow tract and no significant aortic insufficiency. When compared to pre-operatively, normalization of LV dimensions and function was observed postoperatively and at latest follow-up: LV end-diastolic dimension (mean z-scores: -3.4, 0.9 and -0.8 at the 3 time points, respectively), LV end-systolic dimension (mean z-scores: -3.6, 1.4 and 0.1), and LV ejection fraction (mean 76%, 62% and 56%). RV function improved post-operatively for fractional area change (34%, 42% and 48%) and TAPSE (6.9mm, 10.5mm and 15.7mm).
Conclusions: Aortic root translocation was performed in patients with complex TGA/VSD/PS with no mortality and without LVOT reinterventions during follow-up. Most reinterventions involved the RV-PA connection, with a trend towards fewer reinterventions in patients with a valveless RV-PA connection. Ventricular function and dimensions all normalized postoperatively and remained stable during follow-up.

P-264
Safety of oral chloral hydrate as sedative during cerebral MRI in neonates with critical congenital heart disease
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Introduction: To obtain a good-quality MRI, reduce scan-time and increase comfort of the neonate, short-acting sedation may be necessary. Several studies have shown that chloral hydrate (CH) is safe in neonates. However, neonates with critical congenital heart disease (CHD) might react differently to sedation, because of their changed circulation. Therefore, the aim of this study was to examine the safety of CH as short-acting sedative in neonates with CHD.
Methods: This retrospective, observational cohort study included 35 (near-)term neonates who received CH prior to cerebral MRI before and/or after cardiac surgery with cardiopulmonary bypass (surgery:<30 days after birth). Included cardiac anomalies were: single ventricle physiology(N=7), transposition of the great arteries(N=15), aortic arch anomalies(N=9) and pulmonary atresia (N=4). Sedation protocol included oral CH 50-60 mg/kg given by nasogastric tube 15-20 minutes prior to MRI. The following vital parameters were measured directly before and after MRI: heart rate (beats/min), transcutaneous oxygen saturation (%), respiratory rate (breaths/min) and temperature (°C). Number of events with desaturation (>10% decrease) or bradycardia (<100/min) and requirement of respiratory or circulatory support during MRI were reported.
Results: Forty-nine cerebral MRI scans of neonates who received CH prior to MRI were assessed (17 preoperatively and 32 postoperatively). Median dose of CH was 51.6 mg/kg (IQR 50.1-56.3). Saturation and respiratory rate before and after MRI were not significantly different at both preoperative and postoperative timepoint (all: $P>0.05$). Four neonates (8%) had desaturations during MRI (median lowest saturation: 70%). Two patients recovered spontaneously and in the other two patients oxygen support was started during MRI. Heart rate before and after MRI was not significantly different preoperatively ($P=0.072$). Postoperatively, heart rate showed a significant decrease after MRI compared to before ($P=0.003$). In none of the neonates bradycardia was reported and respiratory or circulatory support was needed. After MRI, temperature was decreased when compared to before, both preoperatively ($P=0.006$) and posteroperatively ($P=0.001$).
Conclusion: CH in a dose of 50-60 mg/kg appears to be a safe short-acting procedural sedative in neonates with critical CHD undergoing MRI when appropriate monitoring is applied. Further studies should reveal the efficacy of short-acting sedatives in neonates with CHD.

P-265
Chest drain removal practices in post-operative cardiac patients in paediatric critical care units (PCCU) across United Kingdom (UK)
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Introduction: Chest Drains (CD) are important part of post-op care following paediatric cardiac surgery however the decision making

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

about CD removal, pain assessment and management are variable leading to suboptimal care. Although there is some data on adults, there is insufficient information on paediatric practice in this field.
Objectives: To describe the CD removal practices in post-operative cardiac patients in the PCCUs across UK, especially in relation to analgesia, decision making and post CD removal chest X-ray (CXR).
Methods: A cross-sectional questionnaire survey was sent electronically using Survey Monkey to consultants in all the cardiac PCCUs across UK to get a snapshot of practices in their institutions.
Results:

- The response rate was 90%
- 70% units had a departmental guideline on CD removal
- Decision about CD removal was either solely made by cardiothoracic surgeons (40%) or both PCCU consultants & cardiothoracic surgeons (60%)
- CD was removed by nurses in 80% of the units whereas 10% were removed by PICU doctors and 10% by cardiothoracic surgeons.
- 90% of the units routinely did a CXR post CD removal
- None of the units had a procedural pain score in place for CD removal
- The types of analgesia used by various units included
- Nothing extra 10%
- Morphine bolus 40%
- Ketamine 10%
- Propofol 10%
- Combination of Morphine/entonox/propofol/ketamine or topical agents depending on ventilation status 30%
- The observation period following CD removal was variable ranging from 1-4 hours.

Limitations: Although our response rate was high, this survey has all the inherent limitations related to survey design, small sample size, including only physicians' views.
Conclusions: The survey highlights the variability of CD removal practices in PCCUs across the UK. There is need for reflection on the current practice to remove unwarranted variance and planning further studies in this area with focus on pain assessment/management, patient/parent satisfaction and cost-effectiveness.

P-266
Echocardiographic findings in children with brain death: a one center experience
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Introduction: Brain death is commonly associated with left ventricular systolic dysfunction by some mechanisms not yet completely understood. The aim of this study is to assess echocardiographic features found among organ donors in our hospital.
Methods: This retrospective study was conducted between October 2001 and December 2018. A total of 20 patients under 18 years old with declared brain death were identified. The mean age of the donors was 8.8 years (range 10 months to 17 years), and 14 patients 70)%) were male. Only one patient had previously known cardiac disease, VSD, so his heart was not accepted for heart donation. An adequate transthoracic echocardiogram was obtained in 18 potential organ donors (90%) and all of them were performed before evaluation protocol confirmed brain death.
Results: Echocardiogram was completely normal in 2 patients (10%) and in 11 (55%) patients trace valve regurgitation was found. One patient had moderate mitral and tricuspid regurgitation. Minimal pericardial effusion exhibited 4 (20%) patients. Two patients had mild septal dyskinesia with normal left ventricular ejection fraction.
Global hypokinesia with ejection fraction less than 55% was demonstrated in 5 (37%) and only one patient's ejection fraction was lower than 45%. A total of 14 hearts were harvested for transplantation (70%) after confirming brain death, including a patient with lowest ejection fraction and successfully transplanted.
Conclusion: Mild left ventricular systolic dysfunction occurs often in children with brain death and our overall results suggest that most of these patients could be a heart donor.

P-267
Experience of VV ECMO in children with the Avalon® cannula
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Background: The Avalon® bicaval double lumen cannula (Avalon®) represents an innovative concept for veno-venous(VV)ECMO support in children. We report our experience with the use of this cannula for pediatric respiratory support.
Methods: Retrospective analyses of all VV ECMO support using Avalon® cannula (2014-2018).
Results: Eighteen patients with a median age and weight of 3.6 years (0.1-13) and 19 kg (4.2–50) respectively, received respiratory support using Avalon® cannula. Respiratory failure occurred due to viral or bacterial pneumonia in 7 (including 3 oncologic patients), septicemia in 3, a neoplastic disease in 4, near drowning in 2 and post heart-/chest- surgery in one patient each. VV ECMO was mainly installed at bedside under echocardiographic guidance. In 3 patients, the initial veno-arteriel (VA) ECMO was successfully converted to VV ECMO after a mean of 5 days. In one patient, an initial VV ECMO needed conversion to VA ECMO after 12 days of support; in one patient a second VV ECMO run was needed.
Successful weaning was possible in 16 patients after median support of 6 days (2-32). Two patients died due to their underlying disease. Overall survival to discharge was 83% (n=15). Median ventilation time after VV ECMO withdrawal was 3 days (1-32), median ICU stay 16 days (5 – 64). Three patients suffered 4 major complications (all haemorrhagic): 1 haemopericardium due to cardiac perforation (needing sternotomy); 2 explorative thoracotomy for bleeding after lung biopsy and 1 spontaneous haemopericardium due to therapeutic anticoagulation. Cannula repositioning (3 patients) and change of oxygenator (one patient) were other notable events. 4/16 patients (25%) experienced thrombosis of the internal jugular vein. At a median follow-up of 166 days (0-1400) overall survival was 78%.
Conclusion: VV ECMO with Avalon® cannula provides a safe and versatile respiratory support in paediatric patient population, with excellent survival in reversible lung pathologies. Surgical

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

complications during placement, haemorrhagic complications due to anticoagulation or thrombotic complications of the oxygenator or the internal jugular vein may occur. Once survived to discharge, the long-term survival is good.

P-268

Infective endocarditis in children one decade after change of prophylaxis recommendations

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Background: Infective endocarditis (IE) is a rare but severe cardiac disease in childhood. In 2007 IE prophylaxis recommendations changed. The potential impact of this change was evaluated.
Method: Retrospective data analysis between 2006 and 2017 in a single pediatric heart center regarding diagnostic, clinical, microbial, therapeutic, and outcome variable in children with IE and comparison with the decade before (1996-2005)*.
Results: Twenty-five patients were diagnosed for IE at a median age of 6.91 years (range 0.1 to 19.4, female 48%) with a peak during the first year of life (7/25,28%). All Patients fulfilled the modified Duke criteria for definite (12/25,48%) or probable IE (13/25,52%). The frequency of IE increased from 0.37% in a previous decade* (1996-2005) to 0.59% [n.s.] (calculated as IE cases per hospitalized cardiac patients). Beside two double valve infections, IE affected pulmonary (11/25, 44%), aortic (7/25,28%), mitral (6/25,24%), and tricuspid valve (1/25,4%), ventricular septal defect, patent arterial duct and unknown location (each 1/25,4%), either as postoperative (13/25,52%) and native IE (12/25,48%). Postoperative IE was associated with foreign material (12/13,92%), i.e. prosthetic valves (10/13,77%) or other material (2/13,15%). All (10/10,100%) infected prosthetic valves were pulmonary valves. Predisposing risk factors for IE were found (12/25,48%). Pathogens were staphylococci spp. (8/25,32%), streptococci spp. (7/25,28%), HACEK (3/25,12%), other (4/25,16%) or culture-negative (3/25,12%). Fever (23/25,92%) and reduced clinical status (18/25,72%) were most frequent clinical findings. IE was diagnosed six days after first symptoms (0 to 141). Most frequent complications included new or altered valve dysfunction (14/25,56%) and embolism (8/25,32%). Treatment included antibiotics (25/25,100%), and cardiac surgery (16/25,64%) at 208 (median) days (6 days to 3.13 years), but often early (≤28 days) (9/16,36%) due to valve insufficiency (11/16,44%) or stenosis (6/16,24%), independent of valve localization. Outcome was favourable, beside relapse of IE after 1.47 years (1/25,4%) and mortality due to IE (2/25,8%).
Conclusions: IE is still a severe cardiac disease in childhood, with a constant frequency, relevant morbidity and mortality due to complications, and large rate of re-surgery. The high number of prosthetic pulmonary valve associated IE needs further evaluation and therapeutic alternatives.

*Weber R, et al. *Pediatr Infect Dis J.* 2008;27:544–550.

P-269

Abnormal coagulation factors in single ventricle physiology patients: correlation with hemodynamic parameters and its impact on the postoperative course

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Introduction: Thromboembolic events significantly influence morbidity of patients with univentricular hearts. The presence of an intrinsic or acquired hypercoagulable state has been considered before in these patients. This study evaluates coagulation profiles in a cohort from Norwood stage I through Fontan palliation (stage III) in relationship to liver function, hemodynamic variables and clinical outcome.
Methods: Twenty-six consecutive patients with single ventricle anatomy were included between 2016-2018 and blood samples for assessment of an advanced coagulation profile were taken during cardiac catheterization. Coagulation parameters and invasive preoperative hemodynamic parameters, as well as the clinical course from birth to six months after sampling were retrospectively analyzed.
Results: Mean (±SD) age and weight at time of blood sampling was 76±20 months and 10±4.5 kg, respectively. Ten patients were included before stage II, 13 before and 3 after stage III. Ventricular anatomy was left dominant in 17 patients. Sixteen patients (16/26; 62%) showed decreased antithrombin (AT) and/or protein-C (PC) and/or free-protein-S (PS)-function and/or free-PS-antigen. Two patients showed abnormal activated-protein-C-resistance-ratio due to heterozygous factor-V-Leiden mutation and one heterozygous prothrombin G20210A mutation. Group comparison [abnormal coagulation profile (group 1; n=16) versus normal coagulation profile (group 2; n=10)] showed longer postoperative hospitalization time (p=0.04), longer postoperative catecholamine support (p=0.01), a higher incidence of thromboembolic events (p=0.04) and chylothorax (p=0.007) in group 1. In 5/16 (31%) group 1 patients thromboembolic complications occurred: cerebral stroke (n=1), intestinal ischemia (n=2), thrombus formation in inferior caval vein (n=1) and pulmonary vein (n=1). Extensive, prolonged bleeding was seen in two patients.
Mean pulmonary artery pressure negatively correlated with PC (p=0.02), free-PS-function (p=0.02), and free-PS-antigen (p=0.003). Pulmonary wedge pressure negatively correlated with PC (p=0.03). Oxygen saturation correlated positively (p=0.04) and pulmonary resistance (Rp) negatively (p=0.01) with AT. No correlation was found for anthropometric parameters, stage of palliation, ventricular morphology, end diastolic systemic ventricle pressure and transpulmonary gradient, prothrombin and activated partial thromboplastin time, fibrinogen, liver transaminases, hemoglobin, and platelet count.
Conclusions: Abnormalities in coagulation parameters are common in patients with single ventricle physiology. There is a correlation between hemodynamic and coagulation parameters. Coagulation abnormalities negatively affect postoperative course.

P-270

Single Centre Long-term results of 92 homografts in pulmonary position with 1300 patient years of Follow-up

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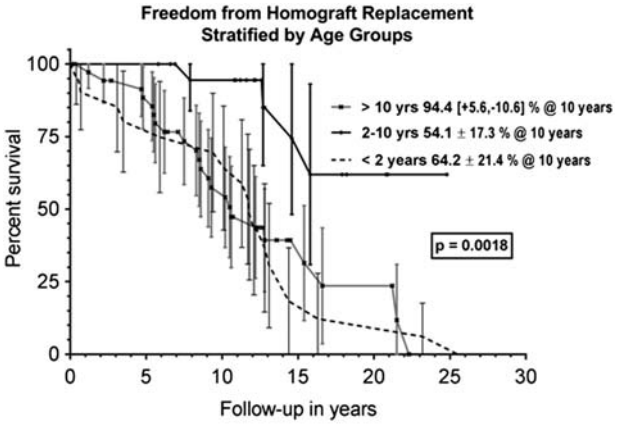
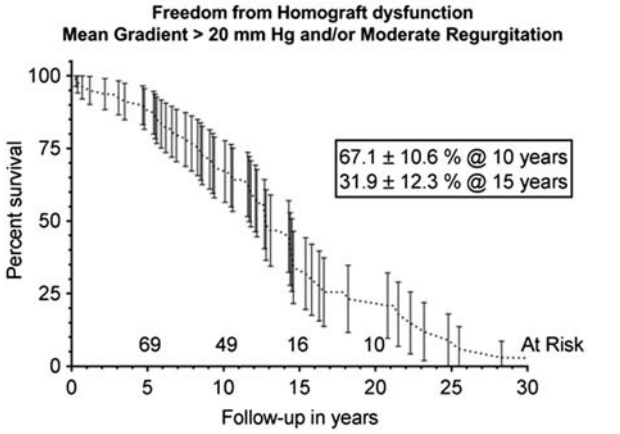
Introduction: To report the long-term outcome of 92 fresh frozen homografts implanted in pulmonary position at a single institution from 1990-2012.
Methods: 92 consecutive fresh frozen homografts implanted were followed up. Median age and weight were 14.3 (0.01-28.7) years and 44.8 (2.5-87) kg respectively. Indications for implantation

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press





were pulmonary valve dysfunction late after primary repair (19), Repair of TOF (17), pulmonary atresia (15), Truncus arteriosus (4), Rastelli Operation (6), Ross Procedure (29) and Miscellaneous (3). Median homograft size was 19.5 (13-26) mm with a Z value of 0.6 (-1.6 to 4.6). Follow-up duration was 14.2 (0.1-28.3) years and was 93% complete.

Results: Kaplan Meier patient survival as well as freedom from endocarditis was 100% at 14 years. Freedom from replacement was 37.4 ± 13.5% at 15 years (At Risk 14). Nine patients were subjected to balloon valvuloplasty for stenosis resulting in freedom from reintervention of 73.9 ± 13.7% at 15 years. Severe graft dilatation/aneurysm formation occurred in two patients. Homograft survival was significantly better in older patients (p=0.0018; Fig 1) and in orthotopic position (p=0.0005). Freedom from composite homograft dysfunction (Mean gradient ≥20 mm Hg and/or Moderate Regurgitation) was 31.9 ± 12.3% at 15 years (Fig 1).

Conclusions: Long-term follow up of homografts in pulmonary position serve as a reference with which the outcome of competing grafts such as Bovine jugular Vein Grafts as well as the newly revisited decellularized homografts would be compared. The incidence of replacement, as well as functional status of the remaining in-situ homografts continues to highlight the need for further innovative approaches to reduce the cyclic need for multiple replacements over a patient's life-time.

P-271
Antibody mediated rejection after paediatric heart transplant

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Introduction: Incidence and clinical significance of antibody-mediated rejection (AMR) after heart transplant (HT) in childhood remains limited due to relatively few paediatric recipients and a lack of routine surveillance.

Methods: All patients under 18 years diagnosed with AMR after HT were included. Routine endomyocardial biopsy (EMB) with C4d and immunohistochemistry were performed in all our patients after HT. AMR severity was assigned using the proposed 2013 ISHLT grading system for pathologic AMR.

Results: A total of 47 patients received a HT between 2008 and August 2018 in our hospital. We performed 311 EMB on them. 25% of the patients (12/47) were diagnosed with AMR, all during the first year after HT. 3 of the 12 patients died: 2 for AMR (6%) and one for noncardiac cause.

All patients had clinical or echocardiographic findings suggestive of rejection. The clinical presentation was low cardiac output (3 patients), right ventricle dysfunction (3 patients, 2 of them requiring ECMO), pure diastolic dysfunction 6 patients.

The grade of AMR was: pAMR (I+) in one patient, pAMR (H+) in one patient, pAMR2 in 8 patients (one died) and pAMR3 in 1 patient (who died). Anti-HLA antibodies were positive only in 2 patients. All patients received treatment with immunoglobulins, 2 with thymoglobulin, 4 with rituximab, 1 bortezomib and 2 with both rituximab and bortezomib. 2 of the 47 HT had positive PRA prior to transplant and received desensitization therapy. None of them developed AMR. Interestingly, six patients had C4D positive with no other evidence of AMR and therefore they were not diagnosed with AMR.

The requirement of mechanical assist devices before HT was not a risk factor for AMR in our patients (p0.5). Nevertheless, the patients who required mechanical circulation after HT had an increased risk of AMR (p0.02).

Conclusions: Regarding our population, DSA does not seem to be very sensible for AMR diagnosis. Clinical and echocardiographic changes are always present when an episode of AMR is recognized. In our patients, the requirement of mechanical circulatory support after but not before HT increases the risk for AMR.

P-272
Is endomyocardial biopsy a reliable tool for diagnosing viral myocarditis in patients with heart failure? Our experience

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Introduction: Acute myocarditis (AM) is an inflammatory disease of the myocardium that can debut as acute heart failure. The differential diagnosis with other entities such as dilated cardiomyopathy (DCM) can be complex. Endomyocardial biopsy (EMB) is considered the confirmatory diagnostic test. Our objective is to review the EMBs of patients affected by AM and compare them with patients with DCM of genetic origin.

Material and methods: We present a descriptive study of the histological and immunohistochemical findings of the EMBs of paediatric patients (0-16 years) diagnosed with AM from July 2007 to Agust 2018. We compare them with the results observed in 4 paediatric patients with DCM with positive genetic test for pathogenic mutations. 6 samples were obtained from the right side of the interventricular septum in all cases. The diagnosis of AM was based on immunohistochemical criteria: 14 or more mononuclear cells and 7 or more CD3 + per mm2.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Results: We detected 42 AM diagnosed patients, 11 by EMB. They presented infiltrate with a median of 25 CD3+/mm2 (13- 80 CD3+/mm2). 6/11 (55%) presented necrosis, 8/11 (72%) fibrosis and 6/11 (55%) edema.
We identified 4 patients with heart failure who underwent EMB with a final diagnosis of DCM of genetic origin. They presented less inflammatory infiltrate (2-24 CD3+/mm2) and 3/4 fulfilled the diagnostic criteria for AM. 1/4(25%) presented necrosis, 2/4(50%), fibrosis and 2/4 (50%) edema.
Hypertrophy of myocardial fibers was more frequent in patients with DCM (75% vs 18%). The viral PCR was positive in 50% of the AM, with good correlation with the plasma PCR. A patient with DCM presented a positive B19V PCR being negative in blood and with suggestive serologies of a past infection.
Conclusions: Although the group of DCM of genetic origin could be affected by AM is improbable and it is more logical to consider that the current criteria are not specific enough. AM is a diagnosis of suspicion based on clinical findings and results of complementary examinations such as echocardiography, magnetic resonance, biochemical and microbiological markers. The immunohistochemical study of the myocardium strengthens the diagnostic suspicion in uncertain cases.

P-273
Risk factors for acute or chronic rejection in pediatric heart transplanted pediatrics

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Objectives: Define characteristics and variables associated with acute and chronic rejection in pediatric patients undergoing cardiac transplantation in our institution.
Methods: Retrospective, observational study. Data of our patients undergoing cardiac transplantation and controlled in our center.
Results: A total 49 cardiac transplants in pediatric patients have been performed in our hospital from 2008. Mean age at transplant was 7,4 +/- 5,9 years old (age range 3 months-17years).
The majority were male 61.2%. Overall, 28.6% (14/49) patients had a diagnosis of congenital heart disease and 69.4% (34/49) patients had diagnosis of cardiomyopathy. Prior to transplant 65.3% (32/49) require some surgical procedure, assistance as a bridge to cardiac transplantation 32.7% (16/49), extracorporeal membrane oxygenation (ECMO) 20,4% (10/49) and 30.6% (14/49) ventricular assist device.
Induction treatment was used in all patients: basiliximab in 61.2% (30/49) and ATG-thymoglobulin in 38.8% (19/49).
ECMO support was required in 18.4% (9/49) cases post-transplant.
Cellular rejection was considered by endomyocardial biopsy in 36.7% (18/49) of the cases, the majority of them in the first-year post transplantation and 26.5% (13/49) as antibodies-mediated rejection (AMR).
There was no statistically significant association between some type of hemodynamic support prior to transplantation (CR p=0,733; AMR p=0,384), diagnosis (CR p=0,715; AMR p=0,547), treatment induction (CR p=0,281; AMR p=1), donor-recipient mismatch (size, sex, Ebstein-Barr and Cytomegalovirus serology), and previous surgical procedures (CR p=0,733, AMR p=0,108) with cellular rejection or AMR. Cardiac allograft vasculopathy has been documented by IVUS in 6.1% (3/49) of patients.
A statistically significant association in patients who required ECMO support for primary graft failure post-heart transplantation and AMR episodes was found (p= 0.001). 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 cardiac support pre-transplantation
Conclusions: Cellular rejection and AMR are not related with clinical features, donor-recipient mismatch (size, sex, Ebstein-Barr and Cytomegalovirus serology) and cardiac support pre-transplant. Children who require ECMO support after cardiac transplantation have greater risk of developing AMR episodes.

P-274
Paediatric cardiomyopathies, integral approach and experience of a tertiary referral centre for paediatric cardiology in Bogotá, Colombia

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Introduction: Cardiomyopathies are a group of diseases of the cardiac muscle with diverse variety of structural and functional phenotypes, being a common cause of heart failure in childhood. Contrary to the adult population, there is a huge knowledge gap regarding the epidemiologic and clinical characteristics of cardiomyopathies in children. This lack of information is even bigger when the situation of medical research in Latin America is taken into account. Given the potential impact that this disorders could have to health-systems, a comprehensive epidemiologic study regarding the clinical profile and sociodemographic characteristics of this subjects, will impact the way we approach paediatric cardiomyopathies.
Methods: An observational, retrospective study was performed over a 7 years period, at a tertiary referral centre for Colombian and Latin American paediatric cardiology. We analyzed all cases of primary cardiomyopathy in children who presented to our institution between 2010-2016 and who were younger than 18-years of age. Cases of cardiomyopathy were classified according to World Health Organization guidelines.
Results: From 29,533 children with suspected heart diseases that attended our institution during the study period, a total of 89 new cases of primary cardiomyopathy were identified. The median age at diagnosis was 11 years. Dilated cardiomyopathy made up 57.3% (N=51) of cases, hypertrophic cardiomyopathy 12.3% (N=11), restrictive cardiomyopathy 8.9% (N=8), non-compacted cardiomyopathy 7.8% (N=7), arrhythmogenic right-ventricular cardiomyopathy 6.7% (N=6) and unspecified cardiomyopathy the remaining 6.7% (N=6). Heart failure was present in 53.93% of patients. The overall mortality was 12.36% (N=11) including 2 of a total of 8 patients who received a cardiac transplantation.
Conclusions: The findings in our study are in agreement with the current scientific literature on the frequency and gender distribution of the different groups of cardiomyopathies in the pediatric age. Currently we do not have any research in Colombia or Latin America that characterizes these patients and it is fundamental to carry out studies like this to understand the magnitude of the problem we face. It is very important to create multidisciplinary groups for the diagnostic and therapeutic approach of these patients, as well as to carry out new studies with a wider population.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

P-275

Impact of preoperative mechanical circulatory support on the postoperative course in heart transplanted pediatric patients

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Introduction: Similar to the adult population an increasing number of children are bridged to heart transplantation (HTx) by the means of various types of mechanical circulatory support (MCS). The use of these devices does not have a negative impact on the long term outcomes in adults, yet relatively limited data are available, especially upon the minor post HTx complications in children.
We reviewed our follow-up data on a comprehensive set of post HTx complications in MCS vs non-MCS population
Patients and methods: 11 MCS patients were included alongside 29 non-MCS subjects. Mean age at HTx was 108 and 101months in the MCS and non-MCS group respectively. Overall mean follow up was 48.95 months (43.6/51 MCS/non-MCS). MCS devices included paracorporeal Berlin Heart biVAD in 3, Berlin Heart LVAD in 3, Levitronics LVAD in further 3, and intracorporeal Heartware in one. One patient was a cross-over form Levitronics to Berlin Heart LVAD
Results: The ground-lying cardiac condition in the MCS group was cardiomyopathy (n=8) and congenital heart disease (n=3). Allosensitisation, which partially responded to desensitization therapy was observed in one assisted case. Overall mortality did not differ between groups (5/29 compared to 4/11 deaths). Clinically and/or histologically relevant rejection, that required intervention differed significantly 5/11 (45.5%), compared to 4/28 (14.2%) cases in the non supported group (p=0.038). Occurrence of HTx related infections, CMV and EBV replication, were comparable in both groups (EBV: 7/29 compared 1/10 cases, CMV: 2/29 compared to 2/11) PTLD was identified exclusively in the non supported group 2/29). Post-HTx diabetes (DM), and autoimmune diseases (AID) were observed in very small number of cases (DM: 2/1 patients in the two groups, AID: 2/1 cases in the two groups)
Conclusions: Similar survival rates can be expected in both bridged and non-bridged paediatric patients following HTx. Transplantation related infections, and other characteristic HTx associated morbidities like diabetes, and AID appeared in similar frequencies. However, the incidence and severity of rejections appears to be more pronounced in the MCS group, which seems to be independent of pretarnsplant allosensitisation.

P-276

Clinical presentation and early predictors for poor outcomes in pediatric myocarditis. A retrospective study

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Introduction: Myocarditis is an important cause of morbidity and mortality in children. The initial diagnosis of myocarditis is usually based on clinical presentation, which is widely variable and in most of cases unspecific. An early recognition is essential in order to monitor and start supportive treatment.
Methods: Retrospective cross-sectional single-group study from January 2008 to November 2017, including children <18 year-old diagnosed as myocarditis. Poor outcome was defined as the occurrence of any of the following: death, heart transplant, persistent left ventricular systolic dysfunction or dilation at hospital discharge (early poor outcome) or after one year of follow-up (late poor outcome). We analysed different clinical features and diagnostic test findings in order to provide some diagnostic clues for myocarditis. Multivariable stepwise logistic regression analysis was performed to determine independent predictor factors of poor early or late outcome.
Results: A total of 42 patients met inclusion criteria. Chest pain (40%) was the most common specific cardiac symptom. Respiratory tract symptoms (38%), shortness of breath (35%), gastrointestinal tract symptoms (33%), and fever (31%) were the most common non-cardiac initial complaints. Signs of heart failure such as heart murmur (26%), systolic hypotension (24%), gallop rhythm (20%) or hepatomegaly (20%) were less prevalent. Up to 43% of patients presented an early poor outcome and 16% presented a late poor outcome.
An initial left ventricular ejection fraction (LVEF) <30% remained the only significant predictor for early (OR (CI95%)=21(2-456); p=0.027) and late (OR (CI95%)=8 (0.56-135); p=0.047) poor outcome. LVEF correlated well with age (r=0.51; p=0.005), days from initial symptoms (r=-0.31; p=0.045), and NT-proBNP levels (r=0.66; p<0.001). NT-proBNP presented a high diagnostic accuracy for LVEF < 30% on echocardiography, with an area under curve of 0.931 (CI95% 0.858-0.995; p<0.001). The best cut-off point was 2000 pg/ml, with a sensitivity of 90%, specificity of 81%, positive predictive value of 60% and negative predictive value of 96%.
Conclusion: Diagnosis of myocarditis in children is challenging. The presence of LVEF<30% on echocardiography at admission resulted the major predictor for poor outcomes. Younger ages, a prolonged course of the disease and NT-proBNP levels could help to identify these high-risk patients.

P-277

Accuracy of NT-proBNP as diagnostic biomarker for incomplete Kawasaki disease among pediatric population

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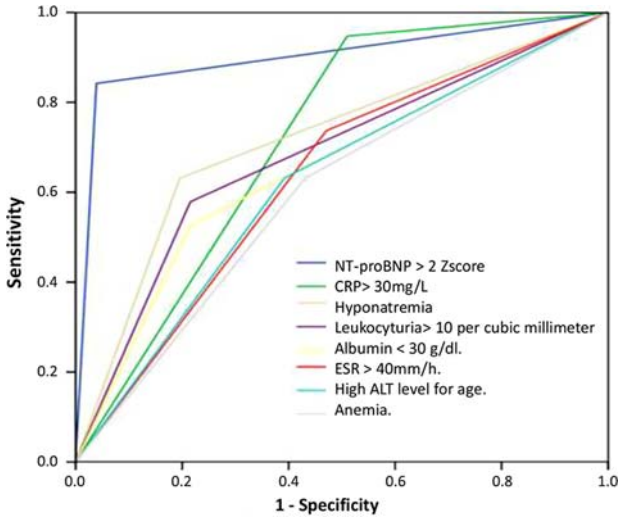
Objectives: The identification of biomarkers for the diagnosis of Incomplete Kawasaki Disease (iKD) is an area of growing research, with NT-proBNP being one of the most promising. We aimed to evaluate the diagnostic accuracy for iKD of plasma NT-proBNP for iKD diagnosis, and to compare it with analitical parameters included in the current AHA`s recommendations.
Methods: We conducted a prospective cohort study including children under 14 year-old admitted with suspected iKD. Patients were divided into iKD and other febrile disease (OFD) based on the final diagnosis made by paediatricians who were unaware of the NT-proBNP value. Through the analysis of ROC curves, the diagnostic accuracy of the analytical parameters of AHA`s recommendations for iKD diagnosis was evaluated and compared respect to NT-proBNP.
Results: During three years, 19 cases of iKD (age 1.42 (0.8-4) years) and 51 cases of OFD (age 3.5 (2-6) years) were included. No differences were found between iKD and OFD in any clinical diagnostic criteria. Patients with iKD had higher CRP, ESR, and NT-proBNP plasma levels than OFD (2424 (1325-3629) pg/mL vs 187 (118-356) pg/mL, p<0.001), and lesser albumin and sodium plasma levels. Of all the parameters analyzed, NT-proBNP showed was best diagnostic accuracy for iKD (Sensitivity 84% and Specificity 96%, AUC 0.901 [0.800-0.987], p<0.001) (Figure1).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



Conclusions: NT-proBNP could be a valid diagnostic test for EKI and potentially superior to the analytical parameters currently recommended in paediatric patients with suspected iKD.

Keywords: NT-proBNP; Kawasaki disease; Biomarkers.

P-280

First use of Ranolazine in adolescent patients affected by hypertrophic cardiomyopathy with refractory symptoms.

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Background: The management of patients with hypertrophic cardiomyopathy (HCM) and refractory symptoms to conventional therapy represents a big challenge for clinical cardiologists. For these patients novel therapeutic approaches are required. However, there is a lack of data in pediatric population and specific trial for evidence of drugs efficacy are still missing. The aim of this study was to report the use of ranolazine in pediatric patients with HCM refractory to standard treatment.
Methods: We collected data of our patients treated with ranolazine before and after the therapy. Data included ECG intervals, NYHA functional class and maximum oxygen uptake (VO2 max) during exercise.
Results: We report 6 cases of adolescent patients (3 males; age range 13-17 years), affected by HCM with refractory symptoms that did not respond to classic therapy. Four patients had restrictive physiology, 3 patients show ventricular dysfunction, 1 patient had obstruction and in 4 out of 5 patients ICD was implanted. All patients were in advanced functional class (NYHA II-IV), and in maximal medical therapy with betablockers and furosemide. The subsequent introduction of ranolazine, based on the experience reported in adults, led to improvement of symptoms, with a reduction in NYHA functional class and an increase in VO2 max at exercise test. No significant changes on ECG were noted.
Conclusion: Introduction of ranolazine in adolescent patients with HCM with refractory symptoms, led to an excellent result with an improvement of NYHA functional class and VO2 max.

P-281

Non-invasive assessment of liver abnormalities in pediatric Fontan patients

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Introduction: Pediatric data about liver abnormalities after Fontan palliation are scarce. We assessed the prevalence and degree of liver problems through non-invasive investigations suitable for longitudinal follow-up.
Methods: 35 Fontan patients (median age 11,8yrs; range 5,2-16,6yrs; 27 boys; median time since Fontan 3,29yrs) were evaluated. A liver ultrasound (US) was performed evaluating nodularity, coarsened echotexture, ascites, liver and spleen size. The diameters of inferior vena cava (IVC), portal vein (PV) in- and expiration and the IVC collapsibility index (IVCCI) were measured. The pulsatility ratio (PR) of the PV and hepatic vein (HV) and the damping index (DI) were calculated. The resistance index (RI) of the PV, hepatic artery (HA) and superior mesenteric artery (SMA) was examined. Fibroscan (Echosens) was used to perform transient elastography (TE). Blood values of AST, ALT, γGT, Alk Phos, bilirubin, total protein, albumin, alpha-foetoprotein, platelet count, cholesterol and Apo-lipoprotein A1 were measured.
Results: Nodularity was found in 2/35 patients and irregularity of the liver surface in 2/35 other patients; hepatomegaly was present in 32% of patients, splenomegaly in 15%. PV mean flow velocity was < 15 cm/sec in 19 (54%) patients, correlating with portal hypertension. 22 patients (63%) showed IVCCI values below 17%, indicative of venous congestion (2). HA RI and SMA RI were inversely correlated with time post Fontan ($p<0,05$; $r2=-0,369$ and $r2=-0,365$ resp.). Liver stiffness was significantly increased compared to controls, with a median(range) of 12,6 kPa (6,6-25,7) versus 4,6 kPa (2-9,5) ($p<0,001$) from early after Fontan. AST, ALT, γGT and direct bilirubin were abnormally increased in respectively 12 (34%), 5 (14%), 24 (69%) and 7 (20%), platelet count was decreased in 7 (20%).
Conclusion: Non-invasive investigations were not able to confirm or differentiate fibrosis from hepatic congestion. We propose follow-up with serial measurements of lab values (ALT, γGT, direct bilirubin, alpha-foetoprotein, platelet count and clotting), US and Doppler parameters (morphology, IVCCI, PV flow velocities, HA RI, SMA RI and PV pulsatility index) and TE. The use of reliable and accurate non-invasive techniques to assess liver fibrosis in children after Fontan remains a major topic for future research.

P-282

Neuregulin-1 improves cardiac function and clinical outcome in a juvenile rat model of right ventricular chronic pressure load

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Introduction: Right ventricular (RV) pressure load (PL) plays a major role in the development of RV failure in congenital heart diseases (CHD). Recently, we revealed an increased proliferative potential in cardiomyocytes of rats with RV PL during childhood. Whether this improves adaptation is unknown. The aim of the

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

present study was to test whether stimulation of proliferation can enhance RV adaptation to PL. We used Neuregulin-1 (NRG1), which is known to enhance proliferation through the ERBB2/4 pathway.
Methods: Rat pups (3 weeks old, 30-40 grams) were subjected to PL by means of pulmonary artery banding or sham at t=0 days. NRG1 or vehicle was administered to rats through intraperitoneal injection from t=3 until t=14 days and sacrificed at t=14, t=28 or t=56 days (young adulthood). We collected clinical symptoms of RV failure, including bodyweight, dyspnea, pleural effusion/ascites and cyanosis. Furthermore we performed analysis of functional and structural adaptation by means of sequential echocardiography and immunohistochemistry.
Results: NRG1 administration in sham animals did not affect hemodynamics or symptoms. Adequate pressure gradient was achieved in all PL rats. Treatment with NRG1 increased cardiomyocyte proliferation (Ki67) after 2 weeks. Treatment with NRG1 in rats with PL markedly postponed the development of clinical signs of RV failure. NRG1 treatment improved cardiac index ($p < 0.05$ at 4 weeks, $p = 0.06$ at 8 weeks).
Conclusions: In the present study we show that NRG1 administration in rats subjected to PL leads to increased cardiomyocyte proliferation during childhood, and this was associated with improved cardiac function and improved clinical outcome. These results support the hypothesis that cardiomyocyte proliferation can be a target as a cardioprotective strategy in children with diseased RV's in CHD.

P-283
Mastering the genetic epidemiology of children with pulmonary arterial hypertension

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Introduction: Pulmonary arterial hypertension (PAH) is called hereditary PAH (HPAH) when the disease is familial or caused by mutations in genes, known to be associated with PAH. In the majority of adult HPAH patients, the mutated gene is bone morphogenetic protein receptor type 2 (BMPR2). In childhood PAH, T-BOX4 (TBX4) and activin-A-receptor-like-type-1 (ACVRL1) mutations seem to be enriched, suggesting a different genetic architecture in children with PAH compared to adults. In addition, children with PAH have been reported to present frequently with associated syndromes or dysmorphic features. The aim of this study is to describe the genetic epidemiology in a national cohort of children with PAH.
Methods: From the Dutch National Network for Pediatric Pulmonary Hypertension (1994-2018) we included 84 children diagnosed with idiopathic PAH, HPAH, PAH associated with congenital heart disease group 3 or 4, or PAH associated with other conditions. Patients were tested on gene mutations in BMPR2, ACVRL1, eukaryotic translation initiation factor 2-alpha kinase 4 (EIF2AK4), Caveolin-1 (CAV1), endoglin (ENG), potassium channel subfamily K member 3 (KCNK3), SMAD9 and TBX4 (with targeted next generation sequencing). Single Nucleotide Polymorphisms (SNP)array for detection of chromosomal numerical variations (trisomies, deletions, duplications) was also performed.
Results: 84 children were enrolled in this study. In 66 children (79%) genetic testing was performed. Absence of genetic testing in 18 children (21%) was mainly due to premature death.
22/66 children had a mutation in a known PAH-associated gene: BMPR2=6, TBX4=7, KCNK3=1, EIF2AK4=1; 2 brothers had PAH associated with a Von Hippel Lindau gene mutation; 5 children had a MMACHC-gene mutation associated with Cobalamin C deficiency and pulmonary veno-occlusive disease. 6/66 children had Down's syndrome. In addition, 20/66 children had genetic variations not known to be associated with PAH. In 18/66 children no genetic variations were found.
Conclusions: Our results show that in 33% and 30% of the study population a PAH-associated gene mutation and additional genetic variations not known to be associated with PAH were present, respectively. The majority of children with PAH (48/66, 73%) have genetic variations.
These pediatric-specific genetic associations might provide clues for the identification of etiologic mechanisms leading to PAH.

P-284
Family form of the restrictive cardiomyopathy: clinical case

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The aim is to optimize the early diagnostics of the restrictive cardiomyopathy, the family form.
Materials and methods: the description of the clinical case of the restrictive cardiomyopathy, the family form.
Results: the child K., 3 years old, was born from the first pregnancy with the risk of pregnancy interruption. The routine prenatal ultrasound examination didn't detect any fetal anomalies. The child arrived on the due date, from the "classic" vaginal delivery. In the family history: the child's grandmother died from the restrictive cardiomyopathy with the atrial fibrillation at the age of 45, the child's uncle died from the restrictive cardiomyopathy at the age of 16. The child's father is now 30 years old. His diagnosis is "Restrictive cardiomyopathy. Atrial fibrillation".
The child underwent echocardiography at the age of 19 days, no abnormalities were found. At the age of 8 months the echocardiography examination showed the left atrial dilatation. At this time the electrocardiography data revealed the both left and right atrial enlargement. At the age of 23 months the child was first hospitalized to the Children's Republican Clinical Hospital with the complaints of small increase in body weight, fatigue, exercise-induced dyspnea. Clinical examination showed paleness of skin integument, stunted physical growth, labored breathing, tachypnea up to 50 per minute, hepatomegaly.
Echocardiography examination showed both left and right atrial dilatation, inferior vena cava and hepatic veins distension, the restrictive form of both left and right ventricles diastolic dysfunction. Electrocardiography examination revealed biatrial enlargement, incomplete right bundle-branch block, ST-T changes. The ultrasound liver examination showed hepatomegaly and hepatic veins dilatation. The NT-proBNP level is significantly increased - 4000 pg/ml. Taking into consideration the family history, the patient was diagnosed with restrictive cardiomyopathy, the family form, NYHA II-III. The standard therapy of heart failure was prescribed. The genetic diagnostics in Moscow Medical Genetic Scientific Centre revealed the mutation $c.4631C > A$ in gene FLNC, mutations in TTN, and the diagnosis was confirmed.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Conclusions: knowing the family history and genetic diagnostics data is incredibly important for the early diagnostics of the family form of the restrictive cardiomyopathy and for the early heart failure treatment.

P-285
Mechanical circulatory support in pediatric age: experience, outcomes and morbidity

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Introduction: Concerning mechanical circulatory support (MCS) in pediatric age, device choice and timing of implantation rely on patient's characteristics and device's risk profile.
The aim of this study is to analyze patient's characteristics, outcomes and morbidity associated to different MCS modalities.
Methods: Retrospective patient data analysis, who needed MCS between 2002 and 2018.
Results: Between 2002 and 2018, 20 patients needed MCS and 22 devices were used. Eleven were on ECMO, eight on pulsatile paracorporeal ventricular assist device (PPVAD) and three on paracorporeal continuous flow ventricular assist device (PCFVAD) with magnetic levitated pump system.
Group A (ECMO): medium age of 4 years, minimum weight 3,2Kg. ECMO was initiated for failure to leave bypass after cardiac surgery (4), refractory postoperative arrhythmia (3), cardiogenic shock (4) in dilated cardiomyopathies (DCM) (2) and congenital heart disease (2). Left ventricular decompression was needed in 2. During a mean of 7,5 days in MCS, 36% experienced major bleeding demanding surgical revision, 9% cannula associated thrombus and 9% neurological events. Renal substitution therapy (RST) was needed in 4. Death occurred in 3 patients (27%), upgrading to another form of MCS in 2 and 5 were weaned off (45%).
Group B (PPVAD): 8 DCM patients (pedimacs 1 and 2), medium age 1,4 years, minimum weight 3,5Kg. PPVAD was used as "bridge to transplant" in all. Mean support time was 71,7 days (7 to 125 days) during which 75% experienced major infection, 37,5% major bleeding, 12,5% device malfunction and 37,5% neurological events. Two needed RST. Half were successfully transplanted.
Group C (PCFVAD): 3 DCM patients (pedimacs 1 and 2), medium age 11,3 years, minimum weight 17,7Kg. PCFVAD was used as "bridge to transplant" in all. Mean support time was 14,6 days (11 to 20 days) during which 66,6% had major bleeding and 33,3% had neurologic events. One patient needed RST. All patients were successfully transplanted.
Conclusion: MCS defined as short term, can be used for longer periods with an acceptable risk profile. The main complications are bleeding and thrombosis related events. The outcomes and prognosis are highly influenced by patient's characteristics and status at time of implantation.

P-286
Repeated measurement of a six-minute walk test has no added value to a single measurement in children with chronic dilated cardiomyopathy

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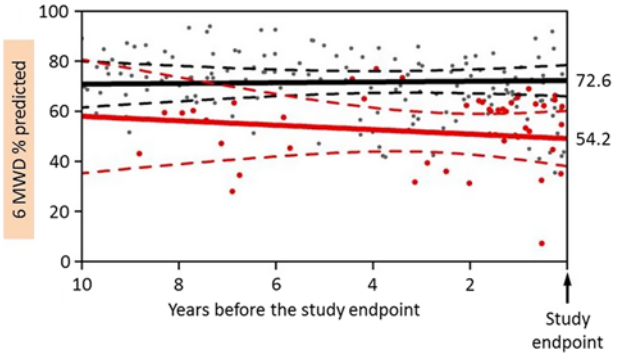

Figure 1.
Serial measurement of 6MDW expressed as percentage of predicted (6MWD%) The average estimates of the longitudinal trajectory of 6MWD%: the black line indicates the patients without a primary endpoint, the red line the patients with a primary endpoint. The dashed lines depict the 95% confidence interval.

Introduction: The six-minute walk test (6MWT) is a feasible test in children with chronic heart failure secondary to dilated cardiomyopathy (DCM) and may be used to identify children with a higher risk of death or heart transplantation. The objective of this study was to determine if repeated measurement of 6MWT has added value in addition to a single measurement in predicting outcome in children with DCM.
Methods: Prospective multi-center cohort study including ambulatory DCM patients ≥ 6 years. A standardized 6MWT was performed at a 6-monthly interval. The distance walked in 6 minutes was expressed as percentage of predicted (6MWD%) based on age and gender specific norm values. We compared change over time in 6MWD% in patients reaching the primary endpoint (composite of death or heart transplantation) to patients who remained endpoint free, using a linear mixed effects (LME) model.
Results: In 57 eligible patients, we obtained n=277 6MWTs, a median of 4 tests (IQR 2-6) per patient during a median of 2.9 years of follow-up (IQR 1.5-5.1). Fourteen patients reached the primary endpoint. Both in patients with and without the primary endpoint 6 MWD% remained fairly constant over time. The mean 6MWD% (based on the LME model) was significantly lower in patients with a primary endpoint than in those without (54% versus 73%; $p<0.001$; see Figure 1). A 6MWD% lower than 63% was associated with an increased risk of heart transplantation or death (hazard ratio 10.8; 95% CI 2.4 to 49).
Conclusions: Children with DCM who died or underwent heart transplantation have systematically reduced 6MWD%. Since their performance is stable throughout follow-up, repeated 6 MWT have little value over a single test within this time window.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Clinical Outcomes of Restrictive Cardiomyopathy in the paediatric population

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Introduction: Restrictive cardiomyopathy (RCM) is rare in childhood. It is characterised by diastolic dysfunction with normal or near normal systolic function, atrial enlargement and normal or near normal left ventricular wall thickness. Diastolic failure leads to congestive heart failure and pulmonary hypertension (PHT). The aim of this study was to review our institutional experience in patients with RCM.
Methods: Retrospective review of all patients diagnosed of RMC between September 2002 and October 2018. Demographic data, mode of presentation, echocardiographic and haemodynamic findings and clinical outcome were evaluated.
Results: 8 children with RCM were identified (4 males, median age 3 years). At the time of diagnosis, 5/8 were considered to have pure RCM and 3/8 RCM/non-compacted cardiomyopathy (LVNC). All the cases were primary cardiomyopathies except one patient with endomyocardial fibrosis due to schistosomiasis.
Mean follow-up was 37.9 months (0.5 - 91). Overall, 6/8 patients developed congestive heart failure and 6/8 PHT. 3/8 patients had arrhythmic events during follow-up: 1 patient had a thromboembolic complication after atrial fibrillation, another developed complete heart block and cardiogenic shock and 1 patient had a sudden cardiac death on the transplant list. All patients with arrhythmias were dead or transplanted at the end of the study.
Out of the all the CT patients with cardiomyopahties in our institution, 5 (14%) were due to RCM. 4 patients received CT and 1 cardiopulmonary transplant. 2 patients required pre-transplant mechanical circulatory assistance (1 Berlin Heart; 1 ECMO and Berlin Heart). 1 patient underwent ECMO post-CT because of RV failure.
Conclusions: Due to the poor outcomes of RCM, early consideration for heart transplant is recommended. Arrhythmias are an important clinical finding in patients with RCM and prophylactic pacing or implantable cardioverter-defibrillator system has to be considered as paediatric patients with RCM are at risk of sudden cardiac death.

P-288

Use of intravenous immunoglobulin in children with inflammatory cardiomyopathy

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Aim: This study is aimed to evaluate the efficacy of intravenous immunoglobulin G (IVIG) as a part of multicomponent therapy of inflammatory cardiomyopathy in children.
Methods: We have enrolled 24 consecutive patients with inflammatory cardiomyopathy, 16 (66,7%) male, 8 (33,3%) female. Mean age 10,1±2,4 (5-17,11) years.
Patients underwent a complete history, physical examination, laboratory studies (including thyroid function, CK, CK-MB, LDG, Troponin I, proBNP, virological and immunologic profile), echocardiography, ECG, Holter monitoring (HM), cardiac MRI, Positron emission tomography (PET) using 18F-fluoro-2-deoxyglucose (FDG). Endomyocardial biopsy was performed according to indications and physician's decision. All patients with inflammatory cardiomyopathy were divided into 2 groups: with decreased left ventricular ejection fraction (LVEF) – 20 (83,3%) and without decreased EF - 4 (16,7%). All were hospitalized with NYHA class I to III heart failure. Cardiac arrhythmias were detected in 10 patients (41,7%).
All patients received IVIG in a total dose of 2 g/kg (0.4 g / kg, N. 5) and standard CHF therapy.
Results: Long-term follow up after the course of IVIG was 19,5±6,1 (6-24) months.
Normalization of the level of CK-MB in 7 out of 15 patients (46.2%) after the treatment.
45% (9/20) patients demonstrated an improvement in LVEF; 45,8% (11/24) decrease in left-ventricular dimensions; 8,3% (2/24) demonstrated the progressive increase of dilatation of the chambers and the decrease in EF, the remaining patients maintained a stable cardiac function parameters. The percentage of patients who improved by at least 1 NYHA class at 6-24 months was 46%.
Among patients with arrhythmias on initial examination, 8 out of 10 showed no cardiac rhythm disturbances during follow-up.
Conclusion: Intravenous Immune Globulin in the Therapy of children with inflammatory cardiomyopathy appears as an effective and safe option in addition to supportive treatment of heart failure.

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Functional capacity in children with Fontan physiology: cardiopulmonary exercise test and echocardiographic parameters

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Cardiopulmonary Exercise Test (CPET) is an essential tool for characterization, prognosis, and response to treatment in children with cavopulmonary physiology (CP-P). Functional capacity decreases with age and the premature detection of impairment is a matter of concern.
This research aimed to describe the functional capacity in children with Fontan physiology, controlled between 2014 and 2017, comparing it with echocardiographic systolic function parameters.
Methods: We reviewed clinical records, echocardiographic and CPET data of 46 children aged 8 - 18 with single ventricle (SV) palliated with Fontan procedure. We described anthropometric measures and physical capacity outcomes. Echocardiographic parameters of ventricular function were differentiated by ventricular morphologies: SV with left morphology (SV-L), SV with right morphology (SV-R), univentricular palliation in biventricular patients named biventricular group (BI-V).
Results: 65% boys; mean age 13.6 ± 2.5 years; mean Body Mass Index Z-Score + 0,3 ± 1. 29 (64.45%) SV-L, 9 (20%) SV-R and 7 (15.55%) BI-V.
Average V'O2peak was 27,3 ± 10,98 ml/Kg/min. Peak Heart Rate was 143 ± 63 bpm, 71% of the expected HR. Peak Respiratory rate was 45 ± 19 bpm, 103% of the expected RR. When analyzing quartiles for V'O2peak, 25% of patients presented values ≤ 23.5 ml/kg/min, which correspond to functional capacity II or lower (NYHA).
Echocardiographic parameters of SV-L patients: 25/29 had EF by Teicholz, 22 (88%) ≥ 55% and 3 (12%) between 54-45%. 19/29 had biplane EF: 16 (84%) had EF ≥ 55% and 3 (16%) between 54-45%. SV-R group: 7/9 were evaluated by FAC, 6 (85.7%)

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

had FAC ≥ 0.33 and 1 (14%) had systolic dysfunction. BI-V patients: 5/7 were evaluated by Teicholz, 3 (60%) had EF ≥ 55%, 1 (20%) between 54-45% and 1 (20%) between 45-30%.
In patients with VO2 less than 24ml/kg/minute no statistically significant relationship was found between low VO2 and systolic or diastolic dysfunction.
Conclusion: 25% of Our cohort of patients with Cavopulmonary physiology showed significantly low VO2/Kg values. Classic echocardiographic parameters did not correlate with these functional alterations. CPET is recommended in the routine control of this patients to have a more reliable assessment of their functional capacity.

P-290
Left ventricular systolic dysfunction in children with chronic kidney disease
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Introduction: Adults with chronic kidney disease (CKD) have an excess of cardiovascular morbidity and mortality, with heart failure (HF) being particularly frequent. Reduced left ventricular ejection fraction (LVEF) defines left ventricular (LV) systolic dysfunction and is associated with poor prognosis. However, CKD patients may have HF symptoms with preserved LVEF. In this subgroup of patients, tissue Doppler imaging (TDI), and two-dimensional speckle tracking echocardiography (2D-STE) can detect LV systolic dysfunction by analysing LV myocardial deformation. The present study evaluated the prevalence of impaired LV global longitudinal strain (GLS) in CKD patients with preserved LVEF and its prognostic consequences. There are limited data in children. The aim of this study was to determine early changes in cardiac function of children with CKD by using TDI and 2D-STE.
Methods: Sixty three children and young adults (57% males, age 14.9±6.4 years) with CKD and preserved LVEF (≥50%) were evaluated. They were compared with age- and gender-matched controls. Left ventricular systolic dysfunction despite preserved LVEF was defined by LV GLS <15%. Peak systolic (s') and early diastolic (e') myocardial velocities in the LV and RV lateral wall, and basal septum were measured at rest using TDI.
Results: All subjects had normal resting LVEF (CKD 65.4±7.6% vs control 67.5±5.7%; p=0.12). However, impaired LV GLS (<15%) despite preserved LVEF was observed in 14% of patients. Overall, CKD patients had lower GLS compared to controls (-17.9±2.8% vs -20.1±1.5%; p<0.001). Moreover, TDI systolic parameters were significantly lower in the CKD group (IVS s' 6.9±1.4cm/s vs 8.6±0.8cm/s; p<0.001) (LV s' 8.5±2.2cm/s vs 10.5±1.7cm/s; p<0.001) (RV s' 10.7±2.2 vs 12.9±1.6cm/s; p<0.001). At rest, LV and RV lateral wall, and basal septal e' velocities were lower in patients (12.2±3.5cm/s vs. 18.5±2.2cm/s, p<0.001; 10.8±3.0cm/s vs. 16.2±2.7cm/s, p<0.001; and 9.8±2.3cm/s vs. 14.6±1.9cm/s, p<0.001; respectively).
Conclusion: The study concluded that TDI and 2D-STE can determine cardiac involvement earlier than conventional echocardiography in children with chronic kidney disease having preserved ejection fraction.

P-291
Diastolic dysfunction is associated with ventricular types and non-administration of vasodilators in patients before Glenn
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Background: Ventricular diastolic dysfunction (DDSV) blemishes pulmonary circulation quite a bit in Fontan circulation whose pulmonary pressures are consisted of venous pressures. We predicted DDSV would adversely affect pulmonary circulation in patients even before Glenn procedure. We investigated cardio-pulmonary factors related to DDSV in patients before Glenn.
Methods: The medical records of 163 pre-Glenn patients were reviewed who were fewer than 2 years. They underwent cardiac catheterization between 2010 and 2018. We defined DDSV as ventricular diastolic pressures 13 mmHg or over. First, we sought related factors to DDSV in pre-Glenn circulation by mono-variate and multi-variate analysis. Second, we inquired into DDSV impacts on pulmonary circulation.
Results: Four factors were significantly different between pre-Glenn patients with and without DDSV. In binominal logistic analysis DDSV was significantly related to following factors: end-systolic ventricular pressure ≥ 91 mmHg (odds ratio 12.3, 95% C.I.: 2.5- 60.7), aortic systolic pressure ≥ 84 mmHg (OR 3.3, 95% C.I.: 1.2- 9.0), and bi-ventricle/left ventricle dominant (OR 2.6, 95% C.I.: 1.2- 5.4), and non-administration of vasodilator/angiotensin receptor blocker (OR 2.1, 95% C.I.: 1.0 – 4.2). The DDSV was not significantly associated with 1st strategies, ventricular volumes, ventricular ejection fraction, and atrio-ventricular valve regurgitation. Out of 4 significant factors, 3 were independently associated with DDSV, such as high end-systolic pressures of ventricle (p=0.0030), non-internal-use ACEI/ARB (p=0.034), non-dominant right ventricle (p=0.019). With regard to adverse against pulmonary circulation, following levels were higher in pre-Glenn patients with DDSV: wedge pressures of pulmonary vein/pulmonary artery pressures (18.5 vs. 14.8 mmHg: p<0.0001); left atrium pressures/wedge pressures of pulmonary artery (8.9 vs. 7.0 mmHg: p=0.0011). The frequency of beta blocker were not significantly different between patients with and without DDSV. However, that of pulmonary vasodilators was significantly higher in DDSV group (20% vs. 6%: p=0.029).
Conclusion: In pre-Glenn patients DDSV were independently associated with high ventricular end-systolic pressures, non-right-ventricle type, and non-administration of ACEI/ARB. Pre-Glenn patients with DDSV possessed highly elevated pulmonary-artery pressures, which would be impeditive to Glenn circulation. We should employ ACEI/ARB more proactively against pre-Glenn patients to acquire well-established Glenn circulation. Especially ACEI/ARB might combat patients with non-right-ventricle type.

P-292
Levosimendan in newborns and infants (perioperative cases excluded) – single centred study.
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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Introduction: Levosimendan is calcium-sensitizing inodilator, which improves cardiomyocytes contractility without increasing oxygen consumption. There is limited experience in the use of levosimendan in children – especially in newborns and infants. Available studies on the use of levosimendan in the pediatric population concern mainly on perioperative treatment, several have endorsed possible benefits of repetitive levosimendan infusion as bridging to heart transplant. This approach is particular important in newborns and infants with refractory ventricular dysfunction, due to shortage of pediatric heart donors, contraindications and higher risks of mechanical assist devices in this group.
Methods: Retrospective analysis of clinical and echocardiographic data, laboratory studies, of all newborns and infants with decompensated heart failure who received levosimendan infusion between December 2016 - October 2018. Exclusion criteria was perioperative levosimendan administration.
Materials: A total of 11 patients were included in the study (9 males - 82%). Diagnosis: severe aortic stenosis – 5 (45,5%); dilated cardiomyopathy (DCM) – 3 (27,2%); HLHS - 2 (18,2%), TGA, LVOTO, VSD - 1 (9,1%). The median age at the levosimendan infusion was 2,5 months (50 days) (range 4 hours – 1 year). Two patients received 2 infusions.
Results: At the moment of levosimendan administration in ICU, 11 patients were receiving complex, conventional treatment without expected response. Tolerance of levosimendan in all patients was good. None of severe adverse effects appeared during the infusions. An overall improvement was observed in 7 cases (63,6%), which led to reduction, then withdrawal of previous inotropic and concomitant treatment, and eventually discharge from ICU. In 4 patients (36,4%) after several days of stabilization, deterioration of general condition had occurred. 1 death was reported due to progression of multi organ failure (DCM patient, 54 days after levosimend infusion).
Conclusion: Levosimendan infusion appears to be effective and safe in significant group of newborns and infants with decompensated heart failure, especially when conventional therapy is not sufficient to maintain stable hemodynamics. However designed studies on the usage of levosimendan on larger pediatric population are necessary.

P-293
Safety and quality of life after a multidisciplinary cardiopulmonary rehabilitation program implementation in children and young adults with complex congenital heart disease and their families

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Introduction and objectives: Cardiac rehabilitation programs (CRP) constitute a potential therapeutic tool for post operated complex congenital heart disease (CHD) patients.
We implemented a multidisciplinary CRP for children and adults with complex CHD. We sought to study its safety and to determine its impact on perception of health and quality of life in our patients and their families.
Methods: 24 patients (13 male), 7 children (8-14yo) and 17 young adults (15-35yo).
CHD included 8 post operated Tetralogy of Fallot, 1 Fontan stage, 8 TGA (4 arterial switch, 4 atrial switch), 3 PAIVS, 2 truncus arteriosus, 2 DORV.
We designed a customized cardiopulmonary rehabilitation program. Variables analyzed included quality of life, psychological tests (SF-36, STAI, BDI, Peds QoL for children and relatives) and ergospirometry. Every week our patients received one psychological session and they completed their tests before and after CRP.
Results: After a mean of 21.5±4 training sessions we observed statistically significant improvement in VO2%predicted ($p<0,05$).
Psychological pre-tests indicated a normal startpoint regarding quality of life, stress and depression, independently of the severity of the disease of our patients. Quality of life questionnaire did not reveal significant changes, which might be explained by study design (excluding highly symptomatic patients for safety with > 50% in NYHA I).
Data revealed that mental dimension of their quality of life was more affected than the physical one. Relatives had a more pronounced perception of the severity of the cardiopathy compaired with their children.
No adverse effects described.
Conclusions: A tailored CRP in children and young adults with complex CHD raised significantly their oxigen consumption, wich is an indirect marker of an improvement in their quality of life.
Our patients described their quality of life, stress and depression indicators as normal, despite the severity of their disease. Their relatives objetified a worse perception.
We propose CRP as a useful and safe tool to improve CHD functional capacity and perception of health.

P-294
Effect of multidisciplinary treatment on Type D personality in obese adolescents and its association with cardiovascular risk

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Introduction: Individuals with Type D personality tend to experience negative emotions (negative affectivity, NA) and to inhibit self-expression in social interaction (social inhibition, SI). Type D has been associated with increased cardiovascular risk in adults and obese adolescents. Obese adolescents display additional cardiovascular risk factors, including vascular stiffness, endothelial dysfunction, and reduced levels of endothelial progenitor cells (EPC). However, little is known about the effect of behavioural intervention on Type D personality characteristics in growing adolescent girls and boys.
Methods: Two cohorts of obese adolescents were recruited: a residentially treated intervention group (n= 33; 15.4 ± 1.5 years, 24 girls and 9 boys; BMI: 36.44 ± 4.82 kg/m^2), receiving supervised diet and exercise training under psychological guidance and an ambulantly treated usual care group (n= 28; 15.1 ± 1.2 years, 22 girls and 6 boys; BMI: 36.72 ± 5.83 kg/m^2). Changes in body mass, cardiorespiratory fitness, microvascular endothelial function and circulating EPC were evaluated after 5 months and at the end of the 10 month program. Established psychological questionnaires were filled in by the participants, including the DS14 measure of Type D.
Results: At baseline scores for NA correlated to vascular stiffness ($p=0.047$) and SI were associated with decreased numbers of EPC ($r=-.39$; $p= 0.04$) independently of BMI.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Residential treatment improved BMI and body fat percentage and increased exercise capacity (p<0.001 after 5 and 10 months). Microvascular endothelial function also improved in the intervention group (p= 0.04 at 10 months). Scores for Type D personality decreased significantly in the intervention group (p 0.004). This was mainly explained by a significant decrease in NA (p=0.037), while the decrease in SI did not reach statistical significance (p=0.057). The association NA and vascular stiffness and between numbers of EPC and SI disappeared after treatment.
Conclusions: In obese adolescents, characteristics of Type D personality are associated with cardiovascular risk factors. The level of Type D personality characteristics in adolescents was reduced by a multidisciplinary treatment program, and the association of Type D with cardiovascular risk factors disappeared after significant weight loss and improvement of exercise tolerance.

P-295

Health Related Quality of Life for School-Age Hypoplastic Left Heart Syndrome Survivors: A Single Center Study

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Introduction: Neurological and radiological research results show an abnormal neurological development in patients treated for hypoplastic left heart syndrome (HLHS). Thus, the aim of this study was (1) to survey the quality of life scores in comparison with healthy children and children with other heart diseases (mild, moderate, and severe heart defects, heart defects in total) and (2) to find out to what extent children with HLHS are successful in integrating socially into daily school life.
Methods: Children with HLHS (ages 6.3 -16.9) under compulsory education requirements who were treated at our clinic between 1997 and 2009 (n=74), were surveyed. 41 children and 44 parents were examined prospectively by psychologists according to Pediatric Quality of Life Inventory, a health-related quality-of-life measurement (HRQOL).
Results: The results of the self-assessments of health-related quality of life (HRQOL) on a scale of 1 – 100 showed a wide range, from a minimum of 5.00 (social functioning) to a maximum of 100 (physical health-related summary scores, emotional functioning, school functioning), with a total score of 98.44. Adolescent HLHS patients rated themselves on the same level as healthy youths and youths with different heart diseases.
Conclusion: The results show that HLHS patients ages 6-16 can be successfully supported and assisted in their psychosocial development even if they show low quality of life scores as well as varying physical and psychosocial parameters.

P-296

Effects of Aortic Arch Surgery on Child Neurological Development

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Objectives: Study the neurophysiological alterations of children who have undergone aortic arch surgery through Selective Cerebral Perfusion.
Methods: Prospective cohort design. The sample consists of children less than 3 years old at the time of repair surgery, with biventricular physiology, operated on aortic arch pathology using cardiopulmonary bypass and SCP, from August 10, 2004 to May 24, 2016. A sample of 81 patients was selected, of whom those who were older than 5 years were evaluated by a child psychologist.
Results: We conducted a study on the patients over 5 years of age (51 boys and girls), where it was observed that at the intelligence level there is not a significant difference with the general population. Perhaps greater lability and dispersion in the results of verbal intelligence is observed. On the other hand, there are clear significant alterations in executive abilities, especially in the capacity to process information (memory and association); however, not as much in the input and output of the information. We detected that this group of children mainly has a delay in the development of auditory memory, visual integration and auditory association. We observed a clear relationship between those children with the diagnosis of ADD and the poor performance in auditory memory tests.
35% were diagnosed with Attention-deficit disorder (ADD) (18 cases out of 51 studied). It is estimated that the prevalence of ADD in the general population of wealthy countries is around 10%.
Conclusions: Patients with an aortic arch pathology operated on during the three year of life using SCP require a neuropsychological follow-up in addition to cardiological, to detect alterations related to learning disorders and to establish treatment of such lesions, if applicable.

P-297

Psychological screening of 8-year-old children with aortic arch obstruction and their families

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Introduction: Children with aortic arch obstruction, particularly those with single-ventricle physiology, have some of the highest rates of neurological injury compared to children with other congenital heart diseases. At school age, approximately one-third of these children manifest cognitive or behavioral problems. Together with the chronic heart condition, these problems pose a burden on parents. Furthermore, parents may feel the need to protect their child and compensate for negative disease related experiences. This can result in a parenting style called compensatory parenting. To explore factors contributing to the development and perseverance of compensatory parenting, we conducted a broad psychological screening of families with a child with aortic arch obstruction.
Methods: We have currently screened 12 eight-year-old children with aortic arch obstructions and their parents. Children were screened for intelligence, executive functioning, emotional and behavioral problems, and quality of life. Parents were screened for psychological problems, quality of the partner relation, sense of control, coping, protective parenting, and perceived vulnerability of the child.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Results: The IQ of the children seems comparable to that of general population, but all children showed executive function problems which hinder their school performance and daily functioning. Furthermore, two-thirds of the children showed behavioral problems which were mainly internalizing. Almost half of the parents had relation problems or were divorced and almost half of the mothers had psychological problems. All parents indicated that they perceived their child as vulnerable and half of the parents were overprotective towards their child.
Conclusions: Eight-year-old children with aortic arch obstruction and their families encounter (neuro)psychological difficulties that have a profound impact on their daily functioning. Structured follow-up is necessary to support these families and prevent escalation of problems by early identification.

P-298
Cardiac problems and palliative care in children with Trisomy 18
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Introduction: There are many controversies concerning type of treatment which should be implemented in children with Edwards syndrome (T18). The aim of our study was evaluation of type of cardiac problems and quality of pediatric palliative home care in children with T18.
Material and methods: The database of prenatal and home care program was used. For quality of home palliative care already describe questionnaire was applied.
Results: Data on 96 fetuses were obtained. There were 13 termination of pregnancy out of 48 diagnosed before 24 weeks (27%), 30 intrauterine deaths, 3 stillbirths and 50 live births. Among the live born: 34 died later in the hospital just after birth, 16 were discharged home under the care of the hospice. 42 children, 17 referred after prenatal diagnosis, were under home based palliative care program. 15 out of 17 (88%) with known T18 after prenatal karyotype were born at time by vaginal delivery, in contrary to 8 out of 25 (32%) without prenatal diagnosis. Birth weight was below normal in all. 38 children had septal defects, majority perimembranous inlet, 30 had primary pulmonary hypertension. One had palliative cardiac surgery. 37 children died at home, 2 at the hospital. 34 died in the first year of life. Time of palliative care lasted between 32 and 1730 days. Three different mechanisms of death were described. The parents highly rated the quality of home palliative care.
Conclusions: Prenatal diagnosis enables decision making about the type of delivery, palliative care and protection of the child against persistent therapy. Neonates born before 33 weeks and weighing <1300 g have no chance to survive. For the first time we proved, that majority of children with T18 have primary pulmonary hypertension, so cardiac surgery for closing the ventricular septal defect should not be performed. Home palliative care is a valuable alternative to hospital treatment in T18 children.

P-299
Asymptomatic hypertrophic cardiomyopathy and elevated troponin levels as first indications of Friedreich ataxia
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Introduction: Friedreich ataxia is a neurodegenerative disease with autosomal recessive heredity. Myocardial involvement usually occurs several years after the onset of the neurological symptomatology in adolescence and early aduldhood.
Purpose: We present three cases with first manifestation of the disease the unusually inflamed myocardium.
Material and Methods: Three children one boy and two girls, aged 10,12 and 11 years old girl underwent pre-athletic screening test by electrocardiography and echocardiography. Because of ECG and echocardiography findings, troponin levels were measured and cardiovascular magnetic resonance (CMR) was performed in all of them. Neurological evaluation and genetic tests followed.
Results: All children were asymptomatic. The ECG showed T wave inversion in leads II, III, AVF, V4-V6 and LV hypertrophy using the Sokolow-Lyon index: SV1+RV6>3.5mV in all of them. Echocardiography showed moderate cardiac concentric hypertrophy, (intraventricular septum thickness/lateral wall thickness, IVS/LA: 12.66±0,57mm/ 12,5±0.7mm) with normal left ventricle without obstruction and normal right and left ventricular function. Measurements of hs-cTnT assay were elevated in all patients vs controls ($p<0.005$). The CMR tissue characterization revealed evidence for oedema, perfusion and fibrosis in all cases. Due to a slightly peculiar walking, neurological assessment was requested and a molecular genetic examination confirmed Friedreich ataxia. Children were referred to a neuromuscular disorders unit.
Conclusion: To conclude a combination of ECG and imaging parameters can reveal early FA-CM and motivate early risk stratification and start of cardiac treatment.

P-300
Clinical Presentation and Diagnostic Approach to Isolated Congenital Coronary Anomalies
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Introduction: Isolated congenital coronary anomalies (ICCA) are rare malformations affecting the pediatric population and can be isolated or associated with other heart diseases. It's clinical presentation varies widely from sudden cardiac death to heart murmur or absence of symptoms, being a casual diagnosis.
The aim of this study is to analyse the initial approach to ICCA. The clinical spectrum is assayed according to each anomaly and the capacity of electrocardiographic and echocardiographic assessment to establish the diagnosis.
Methods: Retrospective monocentric study including the patients under 18 years registered with the diagnosis of ICCA from 1996 to 2017.
Results: ICCA were diagnosed in 27 patients. Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) was the commonest anomaly (41%) followed by right coronaries from left sinus (22%) and fistulas to right ventricle (19%).
The clinical onset with life-threatening symptoms (cardiac failure or cardiac arrest) determines an affection on left coronary artery, the 10 patients admitted due to heart failure being diagnosed of ALCAPA later. Anomalies affecting the right artery are usually clinically silent or appear when studying a heart murmur or chest pain with normal EKG. An association between the presence of a murmur and right ventricle fistulas is described.
At the time of diagnosis, 37% of the patients had electrocardiographic signs of myocardial infarction, appearing in 73% of patients with subsequent ALCAPA.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Echocardiography determined the diagnosis globally in 85% of the cases and 73% of the patients with ALCAPA (left coronary reversal flow being the main echocardiographic marker). Regarding ALCAPA, the diagnostic value of mitral insufficiency, left ventricle dilatation is presented, as well as a decrease in the ejection fraction at diagnosis. In 52% cases, another imaging study was performed to accurate the diagnosis (22% CT scan and 37% coronariography).
Conclusions: A great clinical variety is described, with higher clinical severity in the cases affecting the left coronary artery. The use of echocardiography alone to diagnose coronary anomalies is reliable in most cases. Even so, there are situations, especially in the presence of severe symptomatology, in which performing other techniques to reach the diagnosis is required.

P-301
Usefulness of 3D Printed Models of Congenital Heart Diseases as Educational Tools for Medical Students
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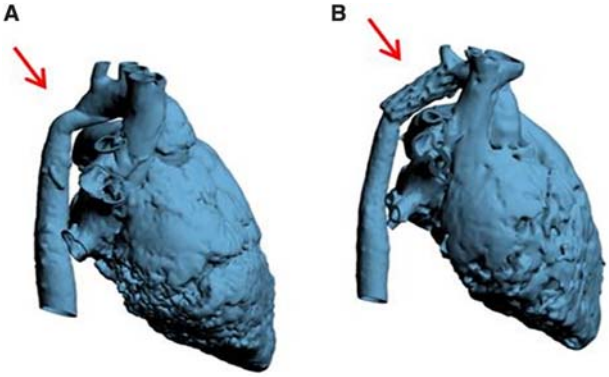
Introduction: Multiple studies have demonstrated the feasibility and accuracy of 3D printed models in the field of congenital heart diseases. These models seem enhance conceptual 3D understanding of complex anatomy. Our aim was to evaluate the usefulness of these models as a teaching tool for medical students to improve learning of congenital heart diseases.
Methods: During the education sessions of left ventricle outflow tract obstruction (LVOTO), students from 5th year of medicine were randomized in two groups. Each group (n=118) attended a 60 minute lecture of LVOTO. All students answered objective questions for pre- and post-lecture knowledge score evaluation, in addition to a subjective post-lecture survey questions regarding students comfort level with the subject. During the lecture, 3D printed models of different types of LVOTO were presented and analyzed by the students in the test group. Knowledge acquisition was evaluated by comparing pre and post-lecture knowledge score. Student's satisfaction and self-efficacy ratings were evaluated by the subjective post-lecture survey. The datas were analyzed and compared between the two groups using paired t-test.
Results: There was no difference in pre-lecture objective test score between the two groups (score 8.32/14 vs. 8.35/14). After the lecture both groups improved their knowledge objective score, but was significantly higher in the test group than that of the control group (score 12.60/14 vs. 11.20/14 respectively p=0.04). Students in the test group were more satisfied with theirs understanding of the diagnosis (p=0.03) and treatment (p<0.01) of LVOTO pathologies.



Conclusions: This preliminary study demonstrates the feasibility and the usefulness of 3D printed models as educational tools of congenital heart diseases for medical students.

P-302
Incidence and prevalence of coronary fistules in the pediatric population
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Introduction and Objectives: to analyze prevalence of coronary fistula in our pediatric cardiology department over a year. Describe the characteristics of these patients.
Methods: retrospective descriptive study. Review of medical records of patients who attended our Pediatric Cardiology Consultation from January 1, 2017 to December 31, 2017, with a diagnosis of coronary fistula.
Results: Out of a total of 2,026 consultations, 30 patients fulfilled criteria for inclusion in the study, which means a prevalence of 0.7 children per 1,000 patients. 46% were women and 54% were men. 26.67% of our patients were born preterm. 43% of the children in the study had problems during delivery or in the neonatal period. The most common type of coronary fistula was the one that drifts into the right ventricle. One patient associated severe complex pulmonary stenosis and perimembranous ventricular septal defect with persistent ductus arteriosus; one ventricular septal defect, Gerbode type; five, foramen ovale; one, aorto pulmonary defect; two, persistent ductus arteriosus and mitral regurgitation. 63.33% of the patients had a family history with heart disease. Two patients are brothers.
Conclusions: The finding of coronary fistulas continues to be an uncommon finding in pediatric patients. The fact that it has increased its prevalence is due to a greater awareness of its existence and the technical improvement of the ultrasound scanners that allow visualizing flows that previously were difficult to diagnose.

P-303
Puffy Layers Of Tummy Points Increased Cardiovascular Risk In Children
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Introduction: Increasing number of kids with puffy layers around their stomach (tummy) directed us to measure waist circumference (WC) as well as body mass index (BMI). In this study, we tried to analyse the metrics of cardiovascular risk factors presence in childhood and relation to the specifically BMI and WC in our local region.
Methods: The study included 152 children ranged 7-11 year. Physical examination included measurements of weight; height, BMI, WC, detailed family history; diet and physical activity were asked with questionnaires'. Variables for the study were total cholesterol (TC), triglycerides (TG), high-density lipoprotein cholesterol (HDL-C), low-density lipoprotein cholesterol (LDL-C), and systolic (SBP) and diastolic (DBP) blood pressure.
Results: We defined SBP < 95th percentile, BMI <85th percentile, WC < 85th percentile, TC < 170 mg/dl, LDL-C <100 mg/dl, HDL-C < 40 mg/dl as a cut-off point, to divide the sample into subgroups with high (risky) and normal values. 17.2% of the subjects were in risky group for SBP, 23.8% had high BMI, 7.3% had high WC, 21.1% high TC, 23.6% low HDL-C and 18.5% high

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

LDL. WC significantly correlated with TG, BMI and inversely correlated with HDL-C blood levels. One% increase in WC decreases HDL-C concentration by 0.99%. One% increase in WC results in 5.35% higher TG. We clustered four cardiovascular risk factors, hypertension SBP>95th percentile, high LDL-C, TC and low HDL-C together in a group. Subjects having at least three of these variables defined as risky cardiovascular group. Children with high BMI were 1.5 times more likely to have clustering cardiovascular risk factors [odds ratio 1.42 (95% CI 0.65–3.2), children with high WC were 2.5 times more likely to have clustering cardiovascular risk factors [odds ratio 2.62 (95% CI 0.81–5.87).
Conclusion: We found the highest percent of reported overweight for children in our region along with high systolic blood pressure. In addition, children with high WC have the highest TG levels. Cardiovascular risk factors could minimized with soft lifestyle changes like going to school on foot, having ideal type 1 diet, the factors that we showed to be protective from CVS risk.

P-304
Cardiovascular disease in a pediatric patient with homozygous familial hypercholesterolemia

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Introduction: Familial homozygous hypercholesterolemia (HFHo) is an autosomal dominant hereditary disease with a prevalence of 1:800.000. To date, 7 are the children diagnosed in Spain. cLDL receptor gene variants are disease causing, resulting in total or almost total lack of enzyme activity. The disease can be diagnosed at birth, showing a total cholesterol (TC) of 700-1000 mg/dl, at the expense of cLDL. Xanthomas, corneal arch, and atherosclerosis are detectable in the first decade of life, while from the second decade an increase of mortality is reported.
Case Report: An 8-year-old boy with multiple tuberous xanthomas and bilateral corneal arch is in actual follow up in our center because his parents are affected by heterozygous familial hypercholesterolemia. His lipid profile at presentation showed TC and cLDL blood levels of 741 mg/dl and 672 mg/dl, respectively, without other biochemical abnormalities. Genetical analysis identified a homozygous rLDL mutation (p.Glu228-stop), confirming the diagnosis. At first, his clinical manifestations comprehended: 1) atheromatous plaques at the carotid vascular US study, 2) a dilated left ventricle (z-score +2.5) at heart US, along with a mild aortic insufficiency and a thickened non-coronary veil, 3) soft atherosclerotic coronary plaques, with no stenosis, and a calcified atheromatous plaque in the ascending aorta at the coronary angioTAC. Ergometry and myocardial perfusion studies were normal. He was started on rosuvastatin and ezetimibe, and also received acetylsalicylic acid prophylaxis. As no improvements where detected, biweekly sessions of LDL-apheresis were also started. A decrease of 50% in baseline cLDL levels, disappearance of tuberous xanthomas, and reduction of carotid atheromatous plaques were observed. Dilatation of the left ventricle persisted. Coronary angioTAC is still pending.
Conclusions / Comments: The initial treatment of HFHo includes dietary recommendations, lifestyle modification and lipid-lowering drugs, but the effectiveness is partial. Our case confirms that LDL-apheresis can be efficient in decrease the cardiovascular risk; we have managed to reduce the skin lesions, as well as atheromatosis and carotid disease. Coronary plaques potential evolution needs more time to be evaluated.

P-305
Cardiopulmonary impact of a multidisciplinary rehabilitation program implementation in children and young adults with complex congenital heart disease

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Introduction and objectives: Cardiac rehabilitation programs (CRP) improve functional capacity in post operated complex congenital heart disease (CHD) patients safely.
We implemented a multidisciplinary CRP for children and adults with complex CHD and their families to quantitatively determine its changes in cardiovascular and respiratory function.
Methods: 24 patients (13 male), 7 children (8-14yo) and 17 young adults (15-35yo).
CHD included 8 post operated Tetralogy of Fallot, 1 Fontan stage, 8 TGA (4 arterial switch, 4 atrial switch), 3 PAIVS, 2 truncus arteriosus, 2 DORV.
We designed a customized cardiopulmonary rehabilitation program including respiratory physiotherapy. We performed EKG, echocardiogram, ergospirometry and 6-minute-walk test.
Results: After a mean of 21.5 ± 4 training sessions, we found statistically significant improvement in forced vital capacity (FVC, +5,6% improvement respect to baseline; p<0,01); maximal inspiratory pressure (MIP, +14,4%; p<0,05); effort time (ET, +12,7%; p<0,01); real metabolic equivalents (METs, +11,3%; p<0,05), VO2%predicted (+3,3%; p<0,05), VO2 at anaerobic threshold (AT, +5,8%; p<0,05) and decreased VE/VCO2 slope (p0,02). 6-minute-walk test (6MWT) mean distance increased from initial 541 ± 94meters to final 642,5 ± 87m (+18,8%; p<0,01).
Echocardiography did not show significant changes.
No adverse effects described.
Conclusions: Tailored CRP are safe and capable to improve cardiorespiratory function in children and young adults with complex CHD. We suggest the implementation of this programs as a therapeutic tool.

	FVC (%)		MIP (cm H_2O)		Effort time (seg)		Direct METs		VO_2 (% predicted)		VO_2 at AT (ml/kg/min)	
	pre	post	pre	post	pre	post	pre	post	pre	post	pre	post
Mean	79	85,1	50,2	56,7	598	650	8	8,4	67,4±	70,5	16,9	18,8
± SD	±15,5	±17,4	± 20,3	± 26,4	±102	±119	±2	±1,7	14	±12,5	±3,7	±4,4
Median	81	85,5	52,8	62	608,5	686	8	8,9	69,2	71,5	17,1	18,1
%change	+5,6%		+17,4%		+12,7%		+11,3%		+3,3%		+5,8%	
p	p < 0,01		p < 0,05				p < 0,05		p< 0,05		p< 0,05	

Forced spirometry, ergometry and ergospirometry statistically significant results. SD standard deviation

P-306
Normal limits for heart rate in children using 24-hour ambulatory electrocardiography

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Background: Knowing the heart rate (HR) appropriate to age is a very important factor for the first screening of cardiac pathology. Several studies provide many patients less than 1 year or adolescents while in our study we analyse HR in all range ages, specially between 3 to 16 years.
Objective: The aim of this study is to establish the normal limits for HR in the paediatric age, using 24-hour ambulatory electrocardiography (AECG), in children without underlying pathology.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Methods: 1211 healthy children, aged between 0 and 18 years, have been examined by AECG. In this study, we specifically analysed 931 subjects with structurally normal hearts and without arrhythmias diagnosed during the AECG recording. Patients were not receiving medication, and did the usual physical activity. Off-line analysis was performed with Mars 8.0 SP3 General Electric. The parameters analysed were age, sex, mean and minimal HR.
Results: All subjects were in sinus rhythm. The subjects were divided by age groups, according to the natural year of birth. We registered patients in all age groups, but most of them were between 3 and 16 years, with at least 20 patients in each group. ANOVA's test showed a significant difference (p <0,01) between HR of each age group. 54% were males and 46% females. The main indications to perform the AECG were: vagal symptoms (31,1%), chest paint (25,1%), palpitations (16,4%). The results for mean and minimal HR were expressed as average and confidence interval for each age group. T-test showed a significant difference between genders, in minimal HR from 9 years and in mean HR from 14 years (p <0,05), being lower in males.
Conclusions: With our study we have shown that mean and minimal HR decrease with age, being more evident from 14 years. We have also proved a significant difference between genders especially during adolescence. This is one of the paediatric studies published so far with the largest number of patients, particularly in school age.

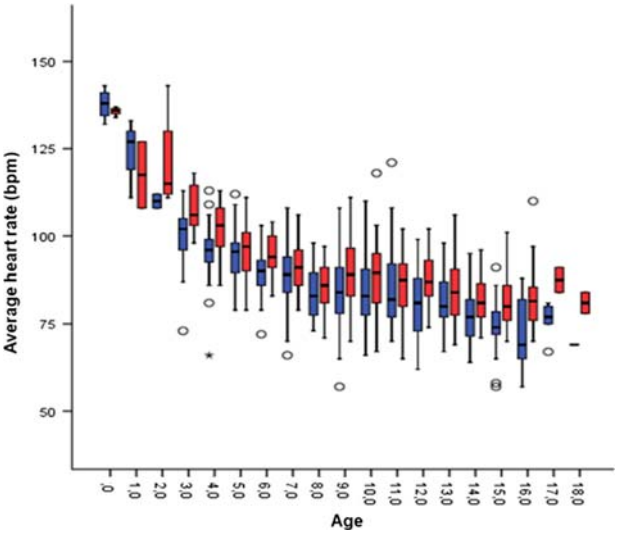

Figure.
Relationship between average HR and age. Blue values represent male sex and red values female sex. Note the decrease in HR with age and difference between genders.

P-307

The etiology and clinical analysis of 232 hospitalized children with hypertension

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Objectives: To analyze the etiology and clinical characteristics of hypertension in children; To analyze the risk factors of primary hypertension with target organ damage.
Methods: We retrospectively analyzed 232 hospitalized children with hypertension in Children's Hospital Insititute of Pediatrics between April 2013 to December 2017.
Results: 1) A total of 232 eligible cases, which consisted of 183 males (79%) and 48 females (21%). Among these children: 38 cases (16%), 126 cases (54%), and 68 cases (29%) were diagnosed as prehypertension, hypertension in stage I, and hypertension in stage II, respectively; 2)The etiological composition of hypertension: There are 181 cases (78%) diagnosed as primary hypertension and 51 cases (22%) diagnosed as secondary hypertension, respectively; The proportion of the primary hypertension children in the period of preschool, school-age, and adolescent was 1.6%, 21% and 77.4%, respectively. The etiology of secondary hypertension mostly are the renal parenchymal hypertension and renal vascular hypertension(43.1%). 3) The clinical manifestations of primary hypertension are mostly asymptomatic or mild symptoms, totally 148 cases(64%); 4) The high risk factors of primary hypertension with target-organ damage including cesarean section, glucose metabolism disturbance and BMI>24kg/m2.
Conclusions: In recent years, the proportion of primary hypertension among hospitalized children has increased. It is of great significance to make blood pressure monitoring as a routine physical examination for primary and middle school students, so as to detect hypertension in children earlier, then make active intervention and improve the prognosis of hypertension in children.

P-308

Home Based Long Term Cardiopulmonary and Inspiratory Muscle Training for Children and Adults with Fontan Circulation

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Objectives: Cardiopulmonary capacity in patients with Fontan circulation is known to be reduced to around 60% of that of healthy individuals (Kempny 2011). Short-term home-based physical training programs have been shown to improve physical function as well as quality of life in these patients (Jacobsen 2016). Inspiratory muscle training has also been shown to improve cardiac output (Laohachai 2017). Aim of this study is to combine these training modalities and show their long-term impact on cardiopulmonary capacity and quality of life.
Methods: Patient above 8 years of age with Fontan circulation were consecutively included in a 24-months training program. They were assessed by reviewing their medical history, routine cardiac examination, cardiopulmonary exercise testing, bodyplethysmography with measurement of inspiratory muscle strength and quality of life questionnaires. The training consisted of home-based bicycle-ergometer training 90 min per week in 3 to 6 training sessions; workload was individually set to 55% of the pre-training maximum values. The second training modality is daily inspiratory muscle training (using a PowerBreathe medic® device). Compliance, heart rate and activity are measured by Fitbit® wrist activity trackers and training journals. Follow-up visits are planned at 4 monthly intervals.
Results: 20 patients were included. After 4 months of training the follow-up data was analyzed: age 10- 43 [mean 20.8] years, 62.5% male, 50% adults. No complications occurred. There were no dropouts. Mean peakVO2 improved from 24.8 to 27.6 ml/min/kg (+2.8ml/min/kg, +14.5%), p=0.029. Mean maximum workload improved from 99.1 to 119.6 Watts (+20.5 Watts, +24.0%),

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

p=0.001. Mean maximum inspiratory pressure (MIP) improved from 6.27 kPa to 8.53 kPa (+2.26 kPa, + 40.9%) p=0.03.
Conclusion: After 4 months of training and having enrolled 20 patients, we can state that the interest in an individual home-based physical training program is high and that home-based training in Fontan patients is safe and shows good compliance. The data shows excellent effects on cardiopulmonary capacity and inspiratory strength.

P-309
Congenital heart disease with hemodynamic repercussion and pulmonary hypertension as predictors of severity in children under five years hospitalized for acute respiratory infection

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Introduction: Acute respiratory infection is one of the main causes of hospitalization in childhood and represents one of the most important causes of mortality in children under 5 years of age. Respiratory complications increase in patients with congenital heart disease, especially in those with hemodynamic repercussion or pulmonary hypertension. The present study was conducted to establish the severity of acute lower respiratory tract infections in children under 5 years of age with congenital heart disease with hemodynamic repercussion or pulmonary hypertension in a pediatric reference hospital in Colombia.
Methods: A cohort-type analytical study was conducted in children under 5 years hospitalized for respiratory infection who underwent an echocardiogram due to a history or suspicion of congenital heart disease or pulmonary hypertension in a pediatric referral hospital in Bogotá between August 2017 and June 2018.
Results: A total of 217 patients were evaluated, of which 62 corresponded to the "exposed" group (congenital heart disease with hemodynamic repercussion or pulmonary hypertension) and 155 to the "unexposed" group. The multivariate analysis found that the exposed group was more likely to enter the PICU (RR 2,269) P <0,001 with a difference of means in days of significant hospitalization (P <0,000). As independent variables of admission to the PICU, infection by respiratory syncytial virus (RR 2.525 P <0.007) and bacterial pneumonia (RR 3.046 P <0.000) were found.
Conclusions: The findings of this study are consistent with those reported in the international literature. The present study showed that children with acute lower respiratory tract infection had a higher risk of admission to the PICU and a longer hospital stay when they coexisted with congenital heart disease with hemodynamic repercussion or pulmonary hypertension. As a predictor of the severity of the respiratory infection, coinfection with respiratory syncytial virus and bacterial pneumonia was also found.

P-310
Hypertension in chronic dialysis pediatric patients

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Introduction: This study aimed to report the frequency, treatment and complications of hypertension in children on chronic dialysis.
Methods: A cross-sectional analysis of all current pediatric maintenance dialysis patients from our tertiary referral center was performed. Data were analyzed using Microsoft Excel and STATA software version 13.
Results: From the 23 patients, sex ratio M:F=1:1.3, mean age 14.1±3.7 years, 12 on hemodialysis, 11 on peritoneal dialysis, mean dialysis duration 30.7±26.8 months, 70% presented hypertension. Some patients had other traditional cardiac risk factors: 2/23 a BMI Z score for age greater than +2 standard deviations and 14/23 dyslipidemia. NT-proNBP was determined and in 48% it had values >4999 pg/mL.
The most used antihypertensive drugs were ACE inhibitors (35%), ARBs (13%), beta blockers (40%), calcium channel blockers (70%) and others like central inhibitors in 13%. In 48% of cases more than one antihypertensive agent was needed.
Most often observed complication was related to the heart: 70% of patients had left ventricular hypertrophy and 48% cardiomyopathy demonstrated on echocardiography but no ischemic event. None had retinopathy or neurological complications.
Conclusions: We found a high prevalence of hypertension in dialysed children. Cardiac disease is the main cause of morbidity and mortality in children with end-stage renal disease and hypertension is the predominant cardiovascular risk factor in pediatric renal patients.
Almost one half of the patients needed more than one antihypertensive drug in order to control blood pressure values.
In the majority of cases cardiac involvement was discovered and almost one half of patients developed cardiomyopathy.
Protocols for regular screening, diagnosis, treatment and monitoring of hypertension are very important in the care of these patients.

P-311
Channelopathies in drowning or near-drowning events in pediatric population

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Swimming or immersion in water can be a trigger for malignant arrhythmias and sudden death in patients with channelopathies (known or concealed) like long QT syndrome (LQTS) or cathecolaminergic polymorphic ventricular tachycardia (CPVT).
Objective: Detection of cardiac channelopathies in patients admitted for drowning or near-drowning in a Pediatric Intensive Care Unit (PICU).
Methods: All cases admitted in the PICU for drowning or near-drowning have been revised. When available, ECG has been analyzed and extended genetic panel has been performed when samples were available.
Results: From 2015 to 2018 our PICU has had 4658 admissions, of which 22 (0,47%) were for drowning or near-drowning events. Fourteen occurred in children that knew how to swim (6-16y). Two died. After comprehensive study including ECG, exercise test and genetic screening, 6/14 were diagnosed of LQTS (4 KCNQ1, 2 negative genetic tests), 2/14 of CPVT (both RYR2+), 1 Brugada syndrome (negative genetic test, drowning occurred during

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

prolonged apnea under water). Five patients had only ECG which was normal, but no further studies were performed.
Conclusion: Drowning and near-drowning events can mask underneath channelopathies. Comprehensive cardiac study should be performed in all cases, including genetic study that in our sample is positive in 66% of the patients.

P-312
Sudden Arrhythmic Death during Exercise: a Post-mortem Genetic Analysis

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Background: Sudden cardiac death is a natural and unexpected death that occurs within one hour of the first symptom. Most sudden cardiac deaths occur during exercise, mostly due to myocardial infarction. After autopsy, some cases, especially in the young, are diagnosed as cardiomyopathies or remain without a conclusive cause of death. In both situations, genetic alterations may explain the arrhythmia.
Objective: Our aim was to identify a genetic predisposition to sudden cardiac death in a cohort of postmortem cases of individuals who died during exercise, structurally normal heart, and classified as arrhythmogenic death.
Methods: We analyzed a cohort of 52 post-mortem samples from individuals <50 years old who had a negative autopsy. Next generation sequencing technology was used to screen genes associated with sudden cardiac death.
Results: Our cohort showed a male prevalence (12:1). Half of deaths occurred in individuals 41–50 years of age. Running was the most common exercise activity during the fatal event, accounting for 46.15% of cases. Genetic analysis identified 83 rare variants in 37 samples (71.15% of all samples). Of all rare variants, 36.14% were classified as deleterious, being present in 53.84% of all cases.
Conclusions: A comprehensive analysis of sudden cardiac death-related genes in individuals who died suddenly while exercising enabled identification of potentially causative variants. However, many genetic variants remain of indeterminate significance, so further work is needed before clinical translation. Nonetheless, comprehensive genetic analysis of individuals who died during exercise enables detection of potentially causative variants and helps identify at-risk relatives.

P-313
Protocol of Cardiac Rehabilitation in Congenital Heart Disease

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Introduction: Cardiac Rehabilitation Programs (CRP) are therapeutic tools recommended by the World Health Organization for over 40 years. In patients with congenital heart disease (CHD) they must be understood as a multidisciplinary project (pediatric cardiologists, rehabilitator, physiotherapist, psychologist and nutritionist). This is our protocol.
Objectives: 1.To ensure the practice of physical activity (PA) in a safe environment and under supervision. 2.To value the cardiac patient in an integral way, evaluating the mental and physical aspect. 3.Reintegrate the child and family into society. 4.Modify risk factors. 5.Improve self-esteem and learn to live with its limitations.
Patients and Methods: Inclusion Criteria: CHD of sufficient severity to restrict the PA of the child, due to either limitations imposed by the children themselves or by their enviroment (doctors, parents, teachers). Ages between 6-17 years. Clinical, electrical and hemodynamic stability. Pathological cardiopulmonary exercise test (CPET) with peak/maximum VO2 <85% predicted. Ability to travel twice a week to the Hospital during the training phase (TP). Signed informed consent. Exclusion criteria: Clinical, electrical or hemodynamic instability. Physical or psychic disability that prevents the performance of the CRP. Documented pathological response to exercise: severe arrhythmias, ST segment alterations, driving blocks with exercise, hypertensive response, hypotension or desaturation <80%. Withdrawal of informed consent.
Place: CR gym (monitoring systems, telemetry and medical cart). Individualized risk stratification according to the results of complementary tests and clinical history. Calculation of the cardiac training frequency (CTF): individualized according to the CPET, incremental limited by symptoms; treadmill; analysis of data with Wasserman method. It will be estimated from the heart rate in the first ventilatory threshold.
Training program: Frequency (2 times/week); Duration (12 weeks, 60 minutes/session); Intensity (CTF and Borg Scale); Specificity (individualized); Phases: recording of constants, warm-up, respiratory physiotherapy, TP (aerobic), cooling. To ensure adherence to the CRP and life habits, two visits will be scheduled at 6 and 12 months.
Conclusion: The CRP in CHD constitute a safe therapy with a positive impact on the quality of life and must be approached in an integral manner, encompassing PA and health education with the aim of better prognosis in medium and long term.

P-314
Disease perception and knowledge about their disease of children with congenital heart defects and their parents

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Introduction: Due to the residuals and sequelae of corrective or palliative surgeries patient with congenital heart disease (CHD) are a chronically ill population. The knowledge of their illness and what it implies, both for children and for adolescents, and for their parents, is a key factor in promoting a good health attitude.
Objective: To evaluate disease perception and knowledge in children, adolescents with CHD and their parents, and whether the degree of understanding is related to age and sex.
Patients and Methods: 85 children 8 years or younger (39,3% girls), 70 from 9-18 years (45% girls) with various CHD and 200 parents participated in this cross sectional study in a Tertiary Pediatric Cardiac Centre over a 6 month period. The questionnaire comprised 15 items that can be grouped to 4 domains: name and nature of heart disease its treatment and prognostic, impact of heart disease on quality of life and daily life, perception of disease and social relationships
Results: 43.8% of patients know the name of their CHD (39.3% ≤8 years, 50% 9-18 years) versus 77.5% parents ($p<0.001$). But 93.3% of the patients have no understanding of their heart defect

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

(≤8 years: 100%, 9-18 years: 85%) compared to 48.5% of the parents (p<0.001). 72.3% of patients believe that they will be cured but only 40% of parents believe that their children will be cured (p<0.001).
52.2% of patients (≤8 years: 15.4%, 8-19 years: 100%) considered their illness not a problem in terms of social relationships compared to 94% of parents (p<0.001).
There was no disagreement in terms of that the children or adolescents with CHD can live the same daily life as their healthy peers (≤8 years: 84.6%, 9-18 years: 89.5%, parents: 79.7%, p=0.45). Also in wellbeing children, adolescents and parents responded similarly to the question "I am (my child) not feeling well" (≤8 years: 7.4%, 8-19 years: 5.3%, parents: 12.9%, p=0.36)
Conclusion: There is a need to intensify CHD-related education to promote a better perception and knowledge of CHD and thus optimize and improve the health behavior of patients.

P-316
Case Report: Symptomatic Inferior Vena Cava Stenosis after Scoliosis Surgery in a Marfan Syndrome Patient with Pectus Excavatum

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Objectives: Marfan syndrome (MFS) is associated with a variety of cardiovascular symptoms, and deformities of the spine and the chest. We report a case of a 15-year-old boy with MFS, presenting with a symptomatic IVC compression after surgical scoliosis correction in the present of an asymmetric funnel chest (FC).
Methods/Results: A 15-year-old Caucasian MFS patient (aortic diameter Z=4.54, systemic score=9 and FBN1-positive) with a severe scoliosis and a FC underwent scoliosis surgery in an outside hospital. During the procedure the patient experienced severe hemodynamic instability, so that he was emergently transferred to our pediatric ICU. On arrival he was found to have severe ascites and extended edema of the lower limbs, which were resistant to diuretics. Imaging findings on a postoperative CT scan suggested a compression of the IVC in proximity to the right ventricle, between the sternum and the spine. Echocardiography demonstrated focal narrowing of the IVC with turbulent flow and a reduced diastolic filling of the right ventricle caused by a severe IVC constriction. Being a well-known patient in our interdisciplinary Marfan center, it was known that a previous MRI had shown a constriction of the IVC on the height of the diaphragmatic passage, which had been clinically asymptomatic at all times - an information that was not available at the outside hospital. Conservative treatment failed to mitigate the disastrous hemodynamic status caused by the inferior inflow congestion. A surgical decompression of the IVC via correction of the FC with a Nuss procedure was performed, leading to a significant improvement of the clinical symptoms. The clinical course proceeded without complications, and the patient was discharged after 3 weeks.
Conclusions: To our knowledge, this is the first case of symptomatic IVC compression in an MFS-patient after surgical correction of scoliosis. In the case of simultaneously existing FC and scoliosis in MFS, the FC correction should be performed before the scoliosis correction to avoid a hemodynamic deterioration. The indication for surgical corrections in MFS patients should be discussed in an interdisciplinary Marfan center, so that the full spectrum of the condition is taken into account.

P-317
N-BNP levels in pediatric Marfan patients - a predictor for left-ventricular pathologies of Ghent Criteria?

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Objective: Correlation of elevated N-BNP levels with reduced diastolic function as well as with dilatation of sinus of Valsalva (SV) was shown before in a collective of pediatric and adult Marfan patients. Isolated data for pediatric Marfan patients is missing and dynamic pathologies of the mitral valve(MV) and its correlation with N-BNP have not been investigated yet. We hereby evaluate correlation of SV dilatation, aortic valve regurgitation (AVR) and MV Marfan pathologies with N-BNP in an isolated pediatric collective. To individualize follow-up regime and therapy our pediatric patients would benefit from early risk stratification.
Methods: Between 2008 and 2018, we diagnosed Marfan syndrome in 171 patients (9.3±5.4y;m:92(54%)) in our specialized pediatric Marfan clinic. In routine follow-up visits, we examined children according to RGC including echocardiography and measured N-BNP with every blood sample. We analyzed N-BNP with age-related standard values 146 times and correlated N-BNP with left ventricular Marfan pathologies (SVdilatation, AVR, MVprolaps, MVregurgitation). In addition, we analyzed N-BNP in 82 patients with medication in comparison to 64 patients without.
Results: N-BNP was not elevated in absence of left ventricular pathologies. Patients with SVdilatation developed higher N-BNP (262±656ng/l) than those without (70±111ng/l;p<0.01). AR patients developed higher N-BNP (376±831ng/l) than patients with competent AV (89±135ng/l,p<0.01). Patients receiving a combined treatment with Sartan and Betablocker(BB) showed significantly higher N-BNP (644±1294ng/l) than patients without treatment (120±79ng/l;p<0.01). In patients treated with a Sartan or BB alone N-BNP levels did not differ significantly in comparison to patients without treatment. MV pathologies did not correlate with higher N-BNP.
Conclusion: As soon as the SV dilates or the aortic valve regurgitates, pediatric Marfan patients´ N-BNP is significantly higher than of those without. But MV pathologies were not associated with N-BNP elevation in our pediatric collective. Thus, unfortunately, N-BNP levels do not gain additional information in pediatric Marfan patients with normal diastolic and systolic function. Nevertheless, it may compliment and affirm existing pathologies like SVdilatation. In conclusion, measurement of N-BNP supplements care of pediatric Marfan patients but regular clinical follow-up examinations including echocardiography and MRI are indispensable and remain gold standard for a safe patient care.

P-318
Feasibility of monitoring physical activity in children using commercial activity trackers

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

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Objectives: Wearable activity trackers are increasingly used in daily life, but most devices are not yet validated for the use in children. They could be used to monitor the activity of children with congenital or acquired heart disease, and the data could be automatically transferred to the pediatric cardiologist. The objective was to assess the feasibility of physical activity tracking in healthy children before and after a standardized surgical intervention using a wearable physical activity tracker.
Methods: This single center, open-label, prospective feasibility study aimed at recruiting 24 healthy children aged 4-16 years undergoing elective tonsillectomy. A physical activity tracker (Withings® Go) was worn by the patients for 10 days before surgery and for 28 days after discharge from hospital. Data and activity diaries were transferred to the study center at the end of each study period. Primary endpoint of this study was the difference in proportions of patients having complete activity measurement data, comparing the tracker with the diary pre- and postoperatively. The study was powered to detect a 35% difference between tracker and diary completeness.
Results: 24 female patients with a median age of 6 (IQR 1) years, a median body weight of 20.5 (IQR 5.8) kg and a BMI percentile of 42 (IQR 55) participated in this trial. For the total duration of the study, 58% of the tracker datasets were complete, vs. only 12.5% of the diaries (difference 45.5%, $p<0.05$), technical failure rate was 29.2%. In the preoperative period, completeness of tracker datasets and diaries was 91.7% and 62.5%, respectively. The tracker data correlated strongly with the parents' estimate during the whole study and after surgery (r2 0.81 and 0.82, respectively, $p<0.01$).
Conclusions: Tracker data and diary records correlated significantly suggesting that trackers could replace diaries in the future. Wearable physical activity trackers might be implemented in the at-home monitoring of children with heart disease and might be a useful tool of assessing the general wellbeing of a child after a therapeutic intervention or to detect clinical worsening of a chronic heart condition during long-term therapy, but still need to be validated in chronically ill children.

P-319
Prescription of exercise to children with univentricular heart and Fontan circulation – a way to retard decline?

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Introduction: after Fontan completion of the univentricular heart cardiopulmonary fitness is reduced and the maximal aerobic capacity typically remains at 43-63% of normal. Often the measurements of lung function are abnormal. The chronically elevated CVP in combination with single pumping systemic ventricle gradually contributes to gradual hemodynamic failure with troublesome life-threatening chronic complications. It is not known how to prevent or postpone deterioration. Habitual exercise of young Fontan patients may have positive effects on peripheral muscle mass and lung function which may enhance Fontan hemodynamics, patient contention and well-being.
Methods: 18 patients with Fontan circulation at 14.6+2.4 years of age (12 boys), 19+2.7 kg/m2 of BMI, and with FM of 18.4+9.5% were recruited. Twelve of the patients had RV- (HLHS) and the rest had LV-systemic ventricle. None of the patients had residual flow through cavoatrial fenestration. The measurements included daily habitual questionnaire, body composition, lung volumetry at rest for forced vital capacity (FVC), forced expiratory volume/sec (FEV1), FEV1/FVC, EUROFIT-testing for comprehensive muscle fitness, and spiroergometry using bicycle. The patients have received an individually tailored exercise prescription for six months after which the measurements will be repeated.
Results: the peak heart rate reached at exercise was 166+15 beats/min, and maxVO2 31.6 + 2.1 ml/kg/min. The patients with RV-systemic ventricle indicated significantly less habitual weekly exercise (2.2+1.6 vs. 5.8+3.h/week, $p<0.005$), had lower limb muscle mass (9,9+1.0 vs. 11.3+1.6 kg/m2, $p<0.03$), lower maxVO2 (27.1+6.0 vs. 31.6+2.1 ml/kg/min, $p<0.05$) without difference in peripheral oxygen saturation (92.4+4.2 vs. 93.1+2.5%, ns.). All patients had restrictive lung function. However, FVC ($p<0.0025$), and FEV1 ($p<0.0001$) were significantly inferior in patients with RV-systemic ventricle without difference in FEV1/FVC suggesting more pronounced restriction of lung function.
Conclusions: our pediatric and adolescent patients with completed Fontan circulation had reduced exercise capacity from normal. The patients with RV-systemic ventricle showed less habitual exercise, and had inferior attributes of cardiopulmonary exercise capacity and lung function. The measures aiming at rehabilitation and maintenance of the patient's physical fitness need to target both ventilatory as well as hemodynamic qualities.

P-320
Preventing coronary artery aneurysms after Kawasaki disease: Who to target for intensified primary therapy? A systematic review and meta-analysis of risk factors

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Introduction: Coronary artery aneurysms (CAAs) after Kawasaki disease (KD) are the leading cause of acquired heart disease in children. Despite prompt treatment with intravenous immunoglobulin (IVIG) plus aspirin (ASA), CAAs still develop in about 5% of patients. Taking into account cost-effectiveness and adverse effects, targeting patients at high risk for CAAs for intensified primary therapy should be considered. Risk factors have been variously reported; hence, we performed a systematic review to inform this strategy.
Methods: We performed a pre-specified search in PubMed spanning 2000-2018, and included studies of children diagnosed with KD and treated with standard IVIG and ASA. We assessed the study designs, quality and heterogeneity, and extracted the risk factors for CAAs as reported as odds ratios (OR) and 95% confidence intervals (CI), where available. We focused on demographic and clinical variables commonly available in clinical practice.
Results: The review included 101 papers. They consist of randomized control trials, prospective and retrospective cohort studies, and case-control studies. A random effects model was used in combination with effect size accounting for heterogeneity regarding study design and parameter definition. Preliminary meta-analysis showed commonly reported risk factors to include age less than 12 months (OR 2.28, 95% CI 1.59 – 3.25), delayed IVIG

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

treatment (OR 3.82, 95% CI 2.38 – 6.13), IVIG resistance (OR 4.42, 95% CI 3.03-6.44), male gender (OR 1.57, 95% CI 1.46 – 1.69), and incomplete KD (OR 4.22, 95% CI 1.09 – 16.26). Higher C-reactive protein (CRP), erythrocyte sediment rate (ESR), platelet and leukocyte count, and low albumin were commonly reported risk factors, with varying cut-off values for prediction of CAAs. We also identified a number of genetic and biomarker studies that reported novel risk factors.
Conclusion: Young age, male sex, incomplete KD, delayed IVIG treatment and IVIG resistance are important risk factors for CAAs after KD, with additional laboratory variables noted. Strategies to identify and target these patients for more intensified primary therapy may further decrease the risk of CAAs. Current strategies targeting IVIG resistance only may be inadequate. Clinical application of novel risk factors may further improve prediction and prevention.

P-321

Obesity in the Paediatric Population – a Risk Factor for Cardiovascular Disorder – 12 Years Case Studies in the North-East Region of Romania

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Introduction: Obesity in paediatric population has a strong impact on all systems and organs, causing metabolic and cardiovascular disorders, both short and long term, significantly increase morbidity and mortality rate. In Romania, the prevalence of obesity in children aged 1-18 years has increased considerably in the last 12 years, ranking it third in Europe.
The purpose of the study is to determine the relationship between obesity and cardiovascular risk in paediatric population, and at the same time, to establish therapeutic management of obesity and cardiovascular disorders.
Methods: In the study, we analyzed 581 children, aged 1-18 years, hospitalized for a period of eleven years (January 2006 – January 2018) in the Paediatric Cardiology Department, "Sfanta Maria" Children's Emergency Hospital of Iasi, Romania.
We observed age, sex, body mass index, blood pressure, biological VSR, serum fibrinogen values, total cholesterol, total lipids, lipidogram and the results of the echocardiography and ophthalmological examination. All paediatric patients in the study group have performed a nutritional study, neuropsychiatric and psychologically exam.
Prospective echocardiographic measurements were performed in 581 obese children. Two-dimensional, M-mode and color M-mode ultrasound, conventional pulse wave Doppler imaging were used to assess cardiac function.
Results: We found increased blood pressure values in 140 of cases. Measurements of LV mass, LV wall thickness and LV end-diastolic diameter and volume were significantly elevated in 101 obese children (17.38% of cases).
VSR, fibrinogen and total cholesterol levels were found to be higher among female subjects (hypercholesterolemia in 16.69% of cases, and 46.12% of cases with increased plasma fibrinogen).
Conclusion: This study showed an increased cardiovascular risk in obese paediatric population in the north east region of Romania The echocardiographic exam confirms the elevated LV mass in obese children.
It is necessary to make a periodic follow-up of height and weight, as well as to evaluate blood pressure, total lipids, lipidogram, total cholesterol, fibrinogen, oftalmological and psychological exam, among children with obesity, in order to prevent cardiovascular complications.

P-322

Variable selection for early diagnosis of congenital heart disease using random forest entropy calculations

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Introduction: Decision trees have been widely used in order to measure the importance of variables. Nevertheless, in the field of bioinformatics their use is not widespread. Populations with high infant mortality need innovative strategies to recognize children at risk.
Methods: Variables present in medical records of patients with congenital heart disease (CHD) have been analysed, searching the most representative signs and symptoms which can lead physicians without experience in paediatric cardiology to recognize newborn and children with congenital heart disease and send them to a reference centre. Combining machine learning techniques and medical knowledge, twenty-eight variables assembled in a formulary based in answers yes/no were chosen as the most representatives for early recognition and diagnosis of cardiac malformation. By the combination of entropy and gain of information as parameters of decision trees it was possible to place variables in a structure according to their importance. The most recurrent signs and symptoms were repeatedly placed as root node of the different trees created for this research, getting as a result very high scores of accuracy in prediction. The use of these variables was backed up by a Pearson correlation matrix, whose results demonstrated the mathematic correlation between all of them.
Results: Entropy values for the variables with the highest correlation with CHD were: Down syndrome (0,99), recurrent pneumonia (0,99), clubbing (0,99), heart murmur (0,98), recurrent low oxygen saturation (0,97) and tachycardia (0,97). Variables with values lower than 0,7 do not help to predict congenital heart disease (e.g. splenomegaly).
Conclusions: This study may provide general practitioners and baseline paediatricians with a validated chart for the early suspicion of CHD and referral to opportune diagnosis and treatment.

P-323

Dobutamine Stress Echocardiography in Early Diagnosis of Cardiac Disease in Childhood Cancer Survivors

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Introduction: Childhood cancer survivors (CCS) have significant risk for cardiovascular morbidity and mortality in part due to anthracycline (AC) cardiomyopathy which is often diagnosed late with poor response to treatment. Early diagnosis is therefore important. Dobutamine stress echocardiography (DSE) is widely used in adult individuals at risk for developing cardiac disease and could be useful for evaluating the degree of subclinical AC-cardiomyopathy in young CCS.
Methods: Eight young CCS (median age 25,1 range 22,9-30,0 years) along with 8 age-matched controls were randomly selected from an ongoing prospective study of early markers for subclinical cardiovascular disease in CCS. All had normal left ventricular ejection fraction (EF) and global longitudinal strain (GLS). DSE was performed using a previously standardized protocol with incremental doses of dobutamine from 5 μg/kg/minute (low phase)

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Table

	Group	Median	Range	P
EF(%)	CCS	61,17	55,9 – 66,4	0,20
	Control	63,35	59,5 – 67,6	
GLS(%) rest	CCS	-21,2	-20,5 – -26,8	0,16
	Control	-22,4	-20,4 - -27,3	
GLS(%) low	CCS	-26,0	-18,4 – -30,0	0,12
	Control	-28,3	-25,6 - -31,4	
GLS(%) max	CCS	-23,6	-17,3 - -28,7	0,06
	Control	-27,8	-22,1 – -30,1	
SSr(1/s) rest	CCS	-0,96	-0,43 - -1,11	0,52
	Control	-0,98	-0,69 – -1,10	
SSr(1/s) low	CCS	-1,79	-1,42 - -2,37	0,89
	Control	-1,80	-1,52 – 2,47	
SSr(1/s) max	CCS	-1,59	-1,34 – 2,45	0,06
	Control	-2,00	-1,51 – 2,24	
SSr-change rest-max(%)	CCS	80,9	42,0 – 135,8	0,03
	Control	107,9	98,9 – 138,8	

to 40μg/kg/min (max phase). GLS (%) and peak systolic strain rate (SSr, 1/s) were calculated from loops acquired in 2-, 3- and 4-chamber views at different phases and mean values were calculated.
Results: Median AC-dose was 160 (97-417) mg/m2. At the maximum heart rate, CCS showed a trend toward lower GLS and SSr (p=0.06) in patients compared to controls. A significant difference between the groups was seen for SSr relative increment from rest to peak stress (p=0.03; Table).
Conclusions: In this relatively small study, young CCS with normal resting systolic function appear to have a tendency for lower GLS and SSr at maximum heart rate in comparison to controls. DSE in CCS might prove helpful in diagnosing AC cardiomyopathy at an early stage.

P-324
Circulating Levels of Endostatin Are Increased in Young Nondiabetic First-Degree Relatives of Patients with Type 1 Diabetes

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Introduction: Previous study at our institution demonstrated dyslipidemic changes in healthy normoglycemic first-degree relatives (FDR) of patients with type 1 diabetes (T1D). In the present study of the same cohort, we aimed to assess circulating levels of novel biomarkers for atherosclerosis and their relationship to the lipid profile and the diabetes-risk HLA DQ2/8 genotype.
Methods: Plasma endostatin, cathepsin S, MMP-9, ICAM-1, VCAM-1 and VEGFr1 were assessed in 70 healthy FDR of patients with T1D (age 12.7±0.6, female: 32) and in 23 age-matched control individuals (age: 11.7±1.0 years; female: 11). Human leukocyte antigen (HLA) genotype 2/8 was assessed in dried blood spots by the DELFIA method. Data are expressed as mean and standard error of the mean.
Results: There was no difference in age, body mass index, arterial blood pressure and C-peptide levels between the FDR and control groups (p>0.2). In the FDR group, plasma endostatin was significantly higher than in controls (p=0.008), whereas cathepsin S, MMP-9 and CAMs-1 plasma levels did not differ between the groups (p>0.3). Endostatin correlated with both total cholesterol and LDL cholesterol (p=0.04, r=0.2 for both) but showed no association with HLA-DQ 2/8 in the FDR group.
Conclusion: To our knowledge, this is the first study indicating elevated levels of endostatin in young normoglycemic individuals at risk for T1D, supporting the hypothesis that early vascular disease may develop before the onset of T1D via mechanisms unrelated to the glycemic levels.

P-325
Carotid stiffness in healthy children and adolescents –reference values in Slovak population

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[Supported by VEGA 1/0044/18]
Introduction: Carotid stiffness represents an important marker of early vascular aging which seems promising for evaluation of the initial atherosclerotic changes and prediction of later cardiovascular events. We aimed to determine the physiological values of carotid stiffness in healthy children and adolescents, which are necessary for interpretation of carotid stiffness values in pathological states and later identification of specific cut-offs for clinical practice. The second aim was to assess the influence of age and sex on carotid stiffness in distinct developmental periods of childhood and adolescence.
Methods: We examined 520 healthy Slovak children aged 7 to 19 years (260 boys) without clinically observed cardiovascular risk factors. Ultrasonography combined with echo-tracking system (Prosound F75 Aloka) on common carotid artery (CCA) was used to analyse the local arterial stiffness. In this 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: The physiological values of carotid stiffness are presented in graphical form for total group, and separately for boys and girls. Index β, Ep, AI and PWV β of the common carotid artery increased with age in healthy Slovak children, while in contrast, AC decreased with age. Statistical analysis did not show significant differences of the evaluated parameters between boys and girls. Correlation analysis revealed the best correlation between four indicators of the CCA elasticity (β, Ep, PWV β, and AC), whereas AI had relatively poor correlation with the other parameters. All the indices of carotid stiffness were dependent on blood pressure at the time of evaluation.
Conclusions: Our study firstly presented the physiological values of the parameters of carotid stiffness for Slovak population of children and adolescents aged 7 to 19 years. All the evaluated parameters (index β, Ep, AI, PWV β, AC) were characterised by significant effect of age, but not sex, during this developmental period. These reference values of carotid stiffness can be used for detection of early atherosclerotic changes in children with various pathological states.

P-326
Interstage-Monitoring of patients with shunt dependent lung perfusion in view of the ventricular type and in context of Interstage-Mortality

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https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Introduction: Interstage-Monitoring is a surveillance program that can help to detect vulnerable patients after shunt implantation. In comparison of patients with univentricular heart of the right and left ventricular type and biventricular heart there might be a difference in the appearance of unexpected complications such as weight faltering, deviations of the desired oxygen saturation and acute shunt occlusion.
Methods: Retrospective analysis of patient data (such as demographic data, data regarding the perioperative period, shunt type, monitoring parameters, monitoring events, laboratory parameters) in 59 patients who had shunt dependent pulmonary perfusion.
Results: Interstage-Monitoring can help to detect signs of shunt complications. There is a specifically vulnerable group in the study population: patients with a univentricular heart of right ventricular type. In comparison to patients with a univentricular heart of left ventricular type or a biventricular heart, these patients had a shorter Interstage-period in total ($p = 0{,}001$, $\alpha = 0{,}05$), since their discharge from the hospital was delayed due to a more complex postoperative course and longer stay on intensive care. There was a significant difference between the groups according to their NT-proBNP levels at the end of the interstage period ($p = 0.038$, $\alpha = 0.05$). Differences of oxygen saturation and body weight were not significant. Mortality occurred only in the group of patients with a univentricular heart of the right type and in the early stage of the Interstage-Monitoring.
Conclusion: Patients with a univentricular heart of the right type have higher complications and mortality risk during the interstage period, but Interstage-Monitoring is an effective surveillance program and can reduce Interstage-Mortality. The relevance of NT-proBNP as a marker for risk stratification needs further investigation but correlates with the risk level.

P-327
Gestational diabetes: Influence of Physical Activity on the mother and her newborn

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Objectives: Gestational diabetes has several effects on the cardiovascular health of the mother and her child. The aim of the study was to measure the impact of physical activity on the health of the mother and her newborn.
Methods: 206 pregnant women were examined in a prospective observational study. A 6-minute-walking-test was performed to evaluate the objective physical activity level. Questionnaires were used to evaluate the daily activities and the physical activities.
Results: Gestational diabetes was found in 99 patients, 107 patients served as healthy controls. Before pregnancy the study group had a significant higher Body -Mass -Index (26,3 vs. 21,6; $p < 0{,}001$) but gained less weight during pregnancy (11,5kg vs 14,9 kg vs., p=0,001).
The objective fitness level was worse in pregnant women with gestational diabetes compared to healthy controls (distance: 472 meters vs. 523 meters, $p = < 0{,}001$). Physical activity before and in pregnancy was less performed in the study group (86% vs. 64,5%, $p = 0{,}002$; 69% vs. 45,7%, $p = 0{,}003$). This corresponds to less daily activity in the study group (walking 69 minutes/day vs. 53 minutes/day, $p = 0{,}144$).
The birth weight in the study group was significantly higher than the control group (p=0,018), although both groups were within normal range. Length and head circumference were equal in both groups.
In the study group, above-average physical activity level resulted in a lower birth weight (3266g vs. 3449g, $p = 0{,}056$) compared to those with a below average activity level in the same group. The birth percentile was significantly lower in the above-average group (36,6 vs. 52,4, $p = 0{,}013$). Length and height didn't show any differences.
Conclusions: Physical activity seems to have a positive additional influence on birth weight in pregnancy with gestational diabetes and might help to optimize treatment. Further interventional studies are needed to prove these results.

P-328
Gestational diabetes: physical activity before pregnancy and its influence on the cardiovascular system

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Objectives: Gestational diabetes mellitus (GDM) is a common complication in pregnancy, affecting around 14% of all pregnancies each year. It will likely further increase, as obesity becomes more and more prevalent. The impact of GDM on cardiovascular changes in pregnant women and her child is still unclear. The aim of the study was to measure the effects of physical activity before pregnancy on the cardiovascular system in patients with gestational diabetes.
Methods: 206 pregnant women were included in this observational study. All participants were recruited in an obstetrical department between 28 - 32 weeks gestation. Questionnaires dealing with pre-pregnancy physical activity were evaluated. The cardiovascular status of the mother included measurements of the intima-media thickness of the carotid arteries (ALOKA prosound 6)
Results: 99 women with gestational diabetes, aged 33,84 (SD ± 4,7) years were examined. 107 healthy pregnant women, aged 32,6 (SD± 4,2) years served as controls. The mean weight in the study group was 73,0 (SD± 20, 3) kg and 61,7 (SD± 9,5) kg in the control group. Based on the higher weight in the study group, the BMI was also significantly higher than in the control group (26,3 (SD± 7,1 vs. 21, 6 ± 3; p=0,001). The frequency of physical activity was significantly higher in the control group ($p = 0{,}001$). Women who were physically inactive before pregnancy had a 3-times higher risk to develop GDM compared to active women (OR = 3,1). The intima media thickness (IMT) of the A. carotis

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

interna was significantly thicker in the study group (0, 47 (SD± 0,004 mm vs 0, 44 (SD± 0,009) mm (p=0,006).
Conclusion: Physical activity and a lower initial weight reduces the risk of developing GDM in pregnancy. Further interventional studies are needed to evaluate these results.

P-329
KidsTUMove wintercamp: A pilot project for children with congenital heart diseases

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Projects accompanying children with congenital heart diseases (CHD) throughout their daily life are important. Winter time very often implements less activity therefore the pilot project kidsTUMove "wintercamp" was established with "kinderherz Südtirol". In the camp, winter activities (e.g. skiing) were accompanied by an interdisciplinary team and tested in a protected field. Through the experience in "wintercamp", it is possible to take part in offers of social life. There is no literature on CHD and its impact at altitude, neither in sports activities nor at altitudes >2000hm.
Methods: 9 children (13, 44 years ±2,46) with CHD participated. To examine the effects from altitude oxygen saturation (SaO2) and reaction time (TDS) was measured at ground level (1000m) and at altitude level of 2200 m. Also SaO2, blood pressure (RR) and pulse was measured pre and post skiing. Health-related quality of life (hQL, KINDL questionnaire) was measured before and after "wintercamp".
Results: Altitude: SaO2 decreased in CHD (p<0,001) and skiing (p < 0.01). Reaction Time increased (p<0.001).
Pre-Post skiing: Children with diseases have significantly lower SaO2 before as well as after skiing (p < 0.01). After skiing RR and pulse were higher than before (p <0.001; p <0.001).
hQL had higher values after the "wintercamp" (87,5 ±6,7, 81,4 ±10,3; p=0.001). Also the physical well-being was higher after the "winter camp" (81, 3 ±11, 7; 74, 3±11; p<0.05).
Conclusion: Since SaO2, RR and pulse are important parameters for daily life activities in CHD and since they have changed in altitude with/without sport activities, theses parameters should be regularly checked in altitudes >2000 hm. Moreover, since the winter camp has an effect in hQL and since it gives an impulse for an active lifestyle during wintertime it is important to provide interdisciplinary prevention offers to CHD (like kidsTUMove). In addition such offers give an opportunity to take part in social activities which are in common in kindergarden, school and families activities also in the season of winter.

P-330
Economic Load of Secondary prophylaxis with Penicillin 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. Secondary prophylaxis with long acting penicillin (Benzathine penicillin G or BPG) injections is the most cost effective method of prevention and control. However poor adherence to BPG has been reported from most high prevalence regions. Inability to afford the treatment may be a contributor to poor adherence rates.
Objective: To study the out of pocket cost of secondary prophylaxis with BPG in pediatric RHD patients.
Methods: We prospectively collected self reported data from the parents of RHD children recruited in the hospital based Pediatric RHD Registry. The data included out of pocket expenses on the drug (BPG), on the transportation to the health facility for injection, and the cost of the provider, for administering injection. For patients interviewed on multiple visits, the mean of all values was calculated. 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) and are converted into Euro (EUR).
Results: The cost data was provided by 547 patients over 1642 visits. The mean age 11.97±3.17 years, 354 (64.7%) were boys. Most patients were from rural areas (91.8%) and from lower and lower/upper lower socio-economic strata on the modified Kuppuswami socio-economic scale (65.6%). The total monthly out of pocket cost of penicillin prophylaxis was INR 60.6±101.5 (EUR 0.75 ±1.25). The drug cost (INR 20±8.46; EUR 0.23±0.10) represented only 31.3% of the total out of pocket cost. Parents spent 44.7% of the total cost on transportation to the health facility and the remainder (23.9%) was spent on administration of the injection (INR 14.5±27.6; EUR 0.18±0.34). The costs of injection provider and the transportation to the health facility exceeded the cost of the drug for 29.6% of the patients.
Conclusions: More than two third of the total economic load of penicillin prophylaxis 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.

P-331
Early nutrition in infants with critical congenital heart defects plays crucial role in their long-term growth

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Introduction: growth failure as a result of feeding difficulties and increased metabolic demands in neonates and infants with critical congenital heart defects (CCHD) may negatively influence their neurodevelopmental outcomes. Moreover, according to so called 'thrifty phenotype' hypothesis a reduced growth in the first 1000 day of life including foetal period may result in a tendency for an obesity and type 2 diabetes if such children are reared in an unhealthy lifestyle in later life. Aims of study was to estimate growth parameters in children with CHD who underwent surgery in the first year of their life.
Methods and Patients: study group consisted of fifty non-syndromic children with CCHD as follows: ventricular septal defect (34%), pulmonary stenosis or atresia (16%), coarctation of the aorta (18%), tetralogy of Fallot (10%) and transposition of great arteries (10%), others (12%).
Anthropometrical data from medical records were retrospectively collected at the moment of the birth and age of six months, three and five years. Using the growth charts for the Czech population from The National Institute of Public Health, the patient's z-scores for height, weight, height-for-age and body mass index were calculated. The stunting was set as z-score for BMI<-2; growth failure as z-score for the height < -2.
Results: The prevalence of stunting was 17% at the birth, 21% at 6 months, 37% at 1 year, 8% at 3 years and 3.6% at 5 years of age.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

The prevalence of the growth failure was 17% at the birth, 11% at 6 months, 13% at 1 year, 16% at 3 years and 25% at 5 years of age.
Conclusions: this study demonstrates a high prevalence of significant malnutrition in infants with CCHS during the first year of life. It was also shown that these children are at increased risk of impaired later growth. As far as growth failure in infancy have an impact on long-term neurodevelopment and quality of life, clinicians have to pay attention to feeding difficulties in infants with CCHD and to standardizing their nutritional practice.
Supported by Ministry of Health, Czech Republic – conceptual development of research organization (FNOL, 0098892).

P-332
Chronotropic incompetence among children and adolescents with congenital heart disease

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Introduction: Impaired exercise capacity is a common feature of congenital heart disease (CHD). In adults with CHD, it has been shown that chronotropic incompetence may contribute to exercise limitation. Systematic data in children and adolescents on this topic is limited. We therefore purposed to assess chronotropic response during treadmill exercise testing in children and adolescents with CHD compared to healthy controls.
Methods: 163 children and adolescents (103 with CHD, median age 15 years and 60 age-matched controls) performed cardiopulmonary exercise testing and were included in this study. Beyond peak oxygen consumption, increase in heart rate from resting level to peak exercise (heart rate reserve) and decrease of heart rate after peak exercise (heart rate recovery) was measured. Chronotropic index was defined as percentage of age predicted maximal heart rate reserve. According to data from adults on bicycle exercise, chronotropic incompetence was assumed for chronotropic index below 0.8.
Results: While resting heart rate was similar between both groups, peak heart rate, heart rate reserve as well as chronotropic index were lower in the CHD group than in controls. Chronotropic index was lowest in patients with single ventricle hemodynamics and correlated with peak oxygen consumption. Heart rate recovery was impaired in the CHD group 1 and 2 minutes after peak exercise compared to controls and correlated with peak oxygen consumption.
Chronotropic index below 0.8 was a relatively frequent finding even in the control group suggesting that the threshold of 0.8 appears inadequate for the identification of chronotropic incompetence using treadmill exercise testing in children. After normalizing to the 2.5th chronotropic index percentile of the control group we obtained a chronotropic incompetence threshold of 0.69.
Conclusions: As an adjunct to measurement of peak oxygen consumption, chronotropic response to exercise appears to be a physiologically important diagnostic parameter in children and adolescents with CHD. However, interpretation of chronotropic response needs to consider age-specific characteristics and the mode of exercise test. Our data may help to interpret future studies on chronotropic incompetence using treadmill ergometer protocols in children and adolescents.

P-333
Arterial Stiffness is Not Yet Increased in Children And Adolescents With Inflammatory Bowel Disease

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Introduction: arterial stiffness increases with the natural aging process. However, it can be accelerated by different factors, including chronic inflammation. This has largely been proved in adult population with various inflammatory diseases including inflammatory bowel disease (IBD), but the data on the initial changes in childhood are still scarce.
The aim of this study was to determine whether there is detectable effect of the inflammation in IBD to the arteries as early as in childhood.
Methods: we used PWVao (aortic pulse wave velocity) measured by Arteriograph (Tensiomed) as a marker of arterial stiffness to detect possible subclinical changes in the arterial wall. We examined the total of 70 children (aged 7 to 18 years -mean 14,39±2,93 years) divided into three groups – patients with active IBD (15 patients), those in clinical remission (35 patients) and healthy controls (20 patients). We used two-tailed t-test for comparisons.
Results: comparing PWVao in patients with IBD (mean 6,15±0,90 m/s) with PWVao in healthy controls (mean 6,02±1,01 m/s), no significant difference was found (p=0,83). Likewise, we found no difference in arterial stiffness comparing children with active disease (mean 6,09±0,74 m/s) and those in the clinical remission (mean 6,21±0,86 m/s) (p=0,50). We came across similar results comparing PWVao in patients with active disease with that in control group. Arterial stiffness was unchanged in patients with Crohn disease (the total of 30 patients) (mean 6,18±0,66) compared to healthy controls (p=0,93). The same was observed comparing PWVao in patients with Ulcerative colitis (the total of 18 patients) (mean 5,94±1,11) and in controls (p=0,44). In two patients the type of IBD is not yet determined.
Conclusions: arterial stiffness in IBD patients at this early age is still unchanged. This suggests that effective IBD treatment in childhood could timely prevent accelerated vascular aging in IBD patients in adulthood.

NO-1
Lack of Knowledge about Contraception and Pregnancy in Adolescent Girls with Congenital Heart Disease – There is Room for Improvement

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Introduction: For adolescent girls with congenital heart disease (CHD), knowledge about contraception, sex and future pregnancies is imperative in order to avoid unplanned high-risk pregnancies, complications related to pregnancies and unnecessary fears. It is essential that pediatric health care providers address these issues during adolescence. They should invite young persons to ask questions and to secure that they receive relevant and adequate information related to their specific CHD.
Objectives: To investigate the knowledge of adolescent girls with a CHD about contraception and the risks during pregnancy.
Method: In this cross-sectional multicenter study, participants from 4 university hospitals in Sweden were recruited. The Knowledge Scale for Adults with Congenitally Malformed Hearts (KnoCoMH) was completed by girls 14-18 years of age with a

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

CHD. Items addressing sex and pregnancy were included in the analysis. Two additional items were added, addressing whether they received any information about contraception and pregnancy and if they discussed future pregnancies in relations to their heart disease during their medical check-ups. Statistical analysis was performed by using Chi-Square test.
Result: In total, 91 girls with a mean age of 15.9 years (±1.1) participated. The distribution of CHD complexity was as follows: mild 36% (n=33), moderate 45% (n=41) or complex 19% (n=17). Only 8% (n=7) stated that they received information about contraceptives in relation to their condition. Similar findings were seen regarding pregnancies, where only 15% (n=13) received information. Girls with moderate and complex CHD (n=13) received information about pregnancies to a higher extent compared to girls with a mild CHD (n=0) (p=0.001). About 64% (n=58) was aware that sexual activity would not aggravate their condition; still 36% was unaware whether sexual activity could worsen their CHD.
Conclusion: Very few participants received information about contraception and pregnancy from their pediatric health care providers. Information and education about contraception, sexual activity and future pregnancy should be provided during adolescence, regardless complexity of the disease to avoid misconceptions and misunderstandings. If needed, adolescent girls should be referred to a specialized gynecologist.

NO-2
Preparing parents for discharge from hospital with their infant after complex cardiac surgery using the Congenital Heart Assessment Tool. An online learning resource for health care professionals

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Introduction: The aim of this session is to present an online learning resource developed within a portfolio of research around the Congenital Heart Assessment Tool (CHAT). The CHAT, an early warning tool for parents to use at home to monitor their infant following the first stage of surgery for complex Congenital Heart Disease (CHD), was developed in 2012 and implemented within a feasibility Study (phase one) at one specialist centre during 2013-2015 (Gaskin, Daniels & Barron 2016; Gaskin Wray & Barron, 2018). Phase two of the project was to evaluate the CHAT in four children's cardiac centres in the UK as part of a Health Improvement Project during 2017 (Smith et al, 2018), resulting in an updated version of the tool (CHAT2).
Methods: This third phase of the project involved development of an online learning resource, which was funded by the University and created in collaboration with Little Hearts Matter, a UK CHD Charity. The aim being to enable wider implementation of CHAT across the UK, through consistent education of health care professionals who are involved in the preparation of parents and families for their infant's discharge.
The learning objectives of the online learning resource are for health care professionals to have:

- Enhanced knowledge and understanding of complex CHD in order to teach parents how to spot signs of clinical deterioration in their infant whilst at home
- Developed an understanding of the CHAT, who it is for, what it does, and why it is used
- Learnt how to use CHAT when teaching parents prior to discharge
- Learnt how to use CHAT to support decision making when taking telephone calls from families at home

The online learning resource recognises different learning styles and incorporates a range of self directed activities, video clips demonstrating how to use CHAT and links to a breadth of supporting resources. The resource is currently being piloted and a staged implementation and evaluation strategy is being planned for 2019. This session will provide an update on the progress of the implementation of this online learning resource, which could subsequently be made available internationally.

NO-3
Retrospective review of running a nurse led Paediatric supraventricular tachycardia clinic

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Introduction: In April 2008 a nurse led supraventricular tachycardia (SVT) clinic was established at Royal Brompton Hospital. The clinic was established to optimise wait times, clinic efficiency and to enhance patient support and experience through clinician continuity. Children seen include those with a structurally normal heart and previously documented SVT. The clinic has been running for ten years, during which time the clinical nurse specialist (CNS) managing the clinic has completed a non-medical prescribing course and an advanced nurse practitioner MSc to support clinical practice and develop professional expertise. The nurse led clinic has four dedicated appointments per week. Support where needed is provided by the paediatric electrophysiology consultants in the concurrently running paediatric arrhythmia clinic. Patients contact the CNS between appointments and in some cases remote management of medications and symptoms can be achieved.
Method: A retrospective audit was performed (April 2008 – December 2017) to identify numbers of patients seen and the outcomes of those patients.
Results:

- A total of 1060 consultations were performed
- Total patients seen was 351
- Diagnoses of patients included; WPW 96 (27.3%), AVRT 72 (20.5%), AVNRT 45 (12.8%), Neonatal SVT 41 (11.7%), Atrial tachycardia 28 (7.9%), SVT (unclassified) 27 (7.7%), Unconfirmed SVT 12 (3.4%), Palpitations 9 (2.6%), PJRT 9 (2.6%), fetal SVT 7 (1.9%), Atrial Flutter 3 (0.85%), Atrial Fibrillation 1 (0.3%), Long RP tachy 1 (0.3%)

The outcomes measured included;

- neonates and infants who were weaned from pharmacological therapy where no further SVT was experienced within the first year of life and who were subsequently discharged
- neonates and infants who were weaned from pharmacological therapy and where no further SVT was experienced within the first year of life but represented with SVT in later childhood
- patients receiving mono, dual or multiple pharmacological therapies
- patients using vagal manoeuvres as a control method
- patients referred for radiofrequency ablation
- patients transitioning to adult arrhythmia services

Conclusion: Nurse led SVT follow up is an effective and safe method of providing care to babies and children with SVT.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Patients benefit from continuity of care during appointments and on-going support between appointments in the management of SVT.

NO-4

Quality of life in Italian Adult Congenital Heart Disease

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Introduction: Advances in modern medicine have led to a noticeable increase in the survival of children born with congenital heart disease (CHD). Today, 85% of such children reach adulthood, and most deaths due to CHD occur after the age of 20 years. In the 2000s, the number of GUCH patients was approximately equal to the number of paediatric CHD patients. In the near future, it is estimated that there will be more GUCH patients than paediatric cases of CHD. Therefore, GUCH patients represent a new population in the health field. The study of GUCH cannot be reduced to only mortality and morbidity indicators but requires extensive investigations. Clinical practice has shown that quality of life (QoL), which is influenced by psychosocial aspects, is often a greater concern than cardiac malformation in GUCH patients.
Methods: A socio-demographic questionnaire and the Short Form-36 scale were administered to a convenience sample of 96 GUCH patients who had undergone surgery for complex CHDs and other CHDs.
Results: The sample (N = 96) contained an equal number of males and females. In the sample, 70% of the participants lived with their family of origin, and just over half had children. In all the dimensions of the SF-36, the patients perceived their QoL as good, regardless of the time of the initial diagnosis of CHD and age. However, age influenced "vitality". The time (duration) since the last surgical intervention influenced the emotional state of the patients,in fact more time passes from the surgical intervention and more low are these such specific scores.
Conclusion: The QoL of the GUCH sample was good, with no differences between the subgroups analyzed. The expectation of a reduced QoL among GUCH patients was not met in this study. This finding may be explained by the young average age of our sample and perhaps also by study methodology. The overall QoL of GUCH patients depends on various psychosocial aspects, and this requires further study using tools other than standardized questionnaires.

NO-5

Transition of adolescents with CHD for adult services – what they want and what healthcare profissionals do

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Introduction: - Adolescents with congenital heart disease transition from pediatric cardiology to adult services should be an intentional and planned process according to their medical, psychosocial and educational needs. The main goal is to promote adolescents independence and autonomy equipping them with skills for decision-making, self-care and self-management of their condition. The objective of this study is to compare adolescent expectations, about the transition process, with the practice of healthcare professionals of reference centres of pediatric cardiology in Portugal.
Methods: – A quantitative, cross-sectional study was conducted. Data was collected in five hospitals, with a questionnaire "Transition of the Adolescent with Congenital Heart Disease for Adult Health Care", "Adolescents" and "Health Professionals" versions, developed for this study. The sample consisted of 93 adolescents and 39 healthcare providers.
Results: - Transference to adult services occurred predominantly (61.53%) between 16 and 17 years and 43.01% of adolescents agree with this age group. However, 30.76% of the adolescents would prefer to be transferred only when they are independent. The predominant reason evoked by health professional (79.48%) for this transfer is age. Health care providers were split about involving the adolescent and the parents in the decision to transfer: 41.02% agree and 30.76% disagree, asserting that the decision is exclusively of the healthcare providers. About 23% of adolescents would not involve the parents in this decision. Healthcare professionals (61.53%) informed that the adolescent meets the adult health team in the first consultation after the transfer, but 23.65% of the adolescents would like to meet the new team at the pediatric cardiology service, previous to transfer.
Conclusions: - Results of this study describe practices of transfer, not transition, to adult health care. When the adolescents reach the pediatric age limit, they are transferred to adult service, according to bureaucratic procedures defined by the institution. Adolescents expectations are not fulfilled in the transition to adult cardiology services.

NO-6

New pain management: new analgesic protocol VS standard analgesia in paediatric cardiac surgery for tardive chest tube removal

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Introduction: Cardiac surgery requires the insertion of drainage into the pleural or mediastinal spaces. Removal of the cardiac/thoracic drainage/tube (Chest Tube Removal: CTR) is associated with moderate to severe pain. These drains are generally removed within 24 to 48 hours after surgery. However, this is not always possible; indeed, in some cases a tube remains inside of a patient for several days and its removal may take place outside of intensive care, in the "general" ward. Pain from CTR can cause inadequate lung expansion; secretion retention due to ineffective cough; and immobility that promotes hypoxemia, atelectasis, and pneumonia. The use of analgesics is the most common method to relieve pain induced by CTR, but this maneuver is often performed without the use of analgesics drugs or other analgesic techniques. Furthermore, the use of specific analgesics outside of environments considered protected, such as intensive care, is still somewhat a taboo. A recent literature review emphasizes how current pain management protocols associated with CTR, using a variety of pharmacological and non-pharmacological techniques, are either non-existent or unsatisfactory. Yet the control of pain is a specific competence and requirement profession—even legally—for all ages.
Methods: A multi-center comparative study that compare two analgesic protocol using 2 different medicines.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

Results: During the period between January 9, 2018 and March 31, 2018, we observed 32 participants in two centers. The average age of participants was five and a half years. T-tests for dependent and independent samples were performed. In the standardized samples, pre- and post-CRT pain evaluations were performed. The pre-procedure evaluation between the two samples shows no statistically significant difference (p=0.154). At post-evaluation, the protocol Group A showed a significant difference (p=0.001) compared to the protocol Group B.
Conclusion: Preliminary results show a greater efficacy with new protocol Group A compared to standard for the management of pain also outside of the pediatric intensive care unit, when CTR is actuated in a ward of pediatric cardiac surgery.

NO-7
Comparative analysis of nursing care during pacemaker implantation, using different approaches of surgery in young children

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Introduction: it is necessary to take into account a number of features during pacemaker implantation in pediatric patients (pts): the age and weight, the dimensions of heart chambers and vessels, the fast growth and high activity level. These are important factors for choosing the lead implantation method and searching for new, more modern and less traumatic approaches. We have developed and evaluated an alternative method: videoassisted thoracoscopic surgery (VATS) implantation of epicardial electrode.
Objective: to compare the nurse support for different kind of surgical approach during pacemaker implantation in children.
Methods: 5 VATS implantations of ventricular epicardial leads were performed in our department. There were 5 pts aged 2 to 4 with (body mass less than 15 kg), with complete atrioventricular block. We evaluated the following data: preoperative care, preparation of thoracoscopic equipment and surgical instruments, anesthetic support time, amount of consumables for the procedure, procedure time, as well as disinfection and all steps of sterilization. We investigated financial and time costs in comparison with the standard endocardial implantation approach and pacemaker implantation using thoracotomy.
Result: Comparative study showed the advantage of the VATS method at the stages of preoperative preparation of the instruments, the anesthesiological and surgical parts, and reduction of financial and time costs. All patients received effective heart stimulation, there were no complications in the postoperative period and all patients leave hospital in 7 days after surgery.
Conclusion: The development of an efficient nursing support algorithm makes it possible to optimize the financial and time costs. VATS procedure in children is less traumatic in comparison with the standard endocardial lead implantation approach and implantation using thoracotomy. We believe VATS to be an appropriate approach for infection risk reducing and postoperative complication prevention.

NP-1
Reactions of Mothers and Fathers after Prenatal or Postnatal Diagnosis of Congenital Heart Disease

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Introduction: A diagnosis of congenital heart disease (CHD) in a fetus or in a newborn triggers increased levels of distress in parents. There is some evidence that mothers' reactions may differ from fathers but this has not been extensively evaluated. The purpose of this study therefore was to investigate reactions of mothers and fathers after a prenatal or postnatal diagnosis of CHD.
Method: Participants were recruited from one university hospital in western Sweden and divided into two groups; Prenatal diagnosis of CHD (Prenatal group) or postnatal diagnosis of CHD (postnatal group). Data was collected at 2-6 months after delivery. The questionnaires used were: The Hospital Anxiety and depressions scale (HAD), Sense of Coherence (SOC-13) and Life satisfaction (LiSat11).
Results: In total 15 parents (7 mothers and 8 fathers) were included in the prenatal group and 30 (15 mothers and 15 fathers) in the postnatal group. The groups were matched regarding age, sex, parity and complexity of CHD. Mothers in the prenatal group scored lower on life satisfaction (Mean 44.4±10.3) compared to mothers in the postnatal group (Mean 56.5±5.2; p=0.013). Fathers in the prenatal group scored higher on depression (Mean 6.25±3.65) compared to fathers in the postnatal group (Mean 3.4±2.69; p=0.04). Furthermore, fathers in the prenatal group scored lower on life satisfaction (Mean 45.4±10.1) and SOC (Mean 56-5±5.1) than fathers in the postnatal group (Mean 56.6±5.1; Mean 75.4±7.0; p=0.0049). No difference was found between mothers and fathers except for a lower SOC in mothers in the postnatal group (p=0.007).
Conclusion: In the prenatal group, both mother and fathers respectively displayed lower life satisfaction compared to mothers and fathers in the postnatal group. Furthermore, fathers in the prenatal group had higher level of depression and lower SOC than fathers in the postnatal group. Few differences were detected within the couple.

NP-2
A Study of ASD management in one UK Centre, comparing length of stay, complication and reintervention

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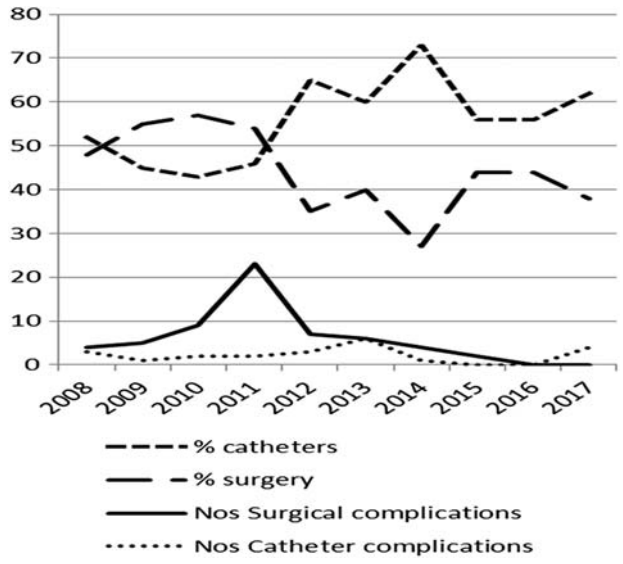
Introduction: Atrial septal defects are diagnosed in one child per 1500 live births, and make up 30-40% of all congenital heart diseases (CHD) seen in adult congenital heart disease (ACHD)1. They are closed with a transluminal device (cardiac catheter), or surgically.
Methods: A retrospective study, between 2008 and 2017, was carried out in one UK centre. The length of stay, reintervention and complication rate was explored for both paediatric and ACHD patients. The percentage of catheter closure compared to surgical closure was studied over the 10 year timeline. The data were collected from the local cardiac database.
Results: The results show a shorter length of stay (LOS) for the patients treated with device closure, median LOS 2 days, versus surgery LOS 7days.
Over the 10 year period, in a cohort of 349 cases managed with catheter closure, 24 had complications, the main complication being failed intervention (8 cases). Paediatric catheter procedure complications were 6/149 (4%), and ACHD complications 16/200 (8%).

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press



In a cohort of 283 cases managed with surgical closure 34 had complications. These consisted of paediatrics: 12/152 (7.9%), and adults 22/131 (16.8%). The main surgical complications were pericardial effusion (10 cases) and arrhythmia (9 cases).
Failed intervention of catheter closure led to all patients having defects closed subsequently.
Of this group, seven patients had surgical closure, and one patient had successful closure via repeat cardiac catheter. The four ACHD patients had longer than average length of stay following surgery (median 11 days) compared to patients who had primary surgical closure.
Over the 10 year period, there was a slow increase in catheter closure of ASD compared to a slow reduction in surgical closure.
Conclusions: Management of closure of ASD is dictated by size and position of the defect, comorbidities, and patient choice. The patients managed by catheter intervention had both a shorter length of stay, and a lower complication rate. Failed intervention of catheter closure led to all patients having defects closed successfully subsequently.
Reference

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NP-3

How to support Hypoplastic Left Heart Syndrome (HLHS) Patients Management during Inter stage

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Introduction: HLHS patients remain as an inpatient following their Norwood Stage I procedure until they have had the Glenn operation. This can mean an admission lasting up to 8 months in total with a long period between the Norwood Stage I and Glenn, referred to as inter stage.
This long stay may be an emotional burden for the families but also to staff caring for the patient during the admission.
The objectives of this project was to gain feedback from staff working on the ward about how they feel about looking after inter-stage HLHS patients.
Methods: Quality improvement methodology (part of a trust leadership programme) was used to access emotional impact on staff of caring for HLHS interphase patients. An emotional touch point model with a non-structured interview was used with two members of staff, and then a short feedback sheet was given to staff nurses, to gain a wider viewpoint on the issue and their thoughts and suggestions for how the situation could be improved
Results: Many suggestions and thoughts were collated from both the emotional touch point exercise, and the feedback sheets. The common themes identified were:
Have a designated medical lead for HLHS patients on the ward during inter-stage
Daily routines/structured meetings
Teaching given to families to allow independence is positive
Teaching for SHO level at medical induction
Nurses felt they were included in planning by registrar/consultant level but dismissed by junior SHO level.
Nurse felt calm and capable, but at times frustrated when they can see a slight deterioration.
Conclusions: Nurses primarily were confident caring for patients during inter stage management, but felt there were improvements to be made in relation to communication and structure.
Improving structure and supporting independence with the families, especially identifying a key medical lead, appeared a positive improvement that could be simply made to current procedures. Also, we will implement an ANP ward round weekly for these families, with emphasis not just on medical stability but also development and weekly achievements.

NP-4

A qualitative exploration of causes of anxiety in parents of children with Tetralogy of Fallot. How can the Cardiac Nurse Specialist team improve current practice?

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Purpose: This service improvement project aims to gain an insight into causes of parental anxiety for parents whose children have Tetralogy of Fallot, and how the cardiac nurse specialist team can improve practice to reduce this stress and anxiety. This study was undertaken at a paediatric cardio-respiratory specialist center in London.
Methodology: for this service improvement project, and Experience-Based Co-Design methodology was utilised. Qualitative research was undertaken in the form of a focus group. Six parents of children diagnosed with Tetralogy of Fallot were asked open ended questions to discuss what caused them anxiety and stress.
Results: The focus group was recorded and data was analysed using thematic mapping. The themes that emerged were; anxiety, communication, quality of life, support in the community and social networks. Once themes were recognised, these themes were analysed and discussed using current relevant supporting literature.
Research Limitations: This research was a single center study in London, therefore this service improvement project is not necessarily representative to other centers or areas of medicine.
Practice Implications: The findings of this project will enable improvements to be made to the cardiac nurse specialist service. A multi-disciplinary team meeting will be held to circulate the results, and plan how we can better support families whose children are diagnosed with Tetralogy of Fallot based on the results found within this research.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press

NP-5

A research protocol on the implementation of the Synergy Model in an Italian Cardiac Surgery Unit: towards a concrete patient centered care

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Introduction: The need to deliver tailored care to meet the peculiarities of the patients' needs is a crucial matter. So far, delivery of care is often based on a fragmentation of tasks and activities. In this scenario, the implementation of a patient-centered care model is important, considering the patients in a holistic way and avoiding the fragmentation of the care delivery. The Synergy Model is a new nursing model nursing that addresses the characteristics of the patients and the nursing competence, showing in literature evidence of positive effects on patient's clinical outcomes and overall satisfaction of patients and nurses.
However, no experiences on the implementation of this model in Italy are available. For this reason, this research protocol aims to assess the effects of the Synergy Model implementation on clinical and psychological outcome in patients from an Italian Cardiac Surgery Unit, as well as analyzing the effects on the healthcare system costs.
Methods: The study is a single-arm longitudinal trial. The data collection will be performed at baseline (before the implementation) and every 4 months (after the implementation), for 3 years. Clinical and psychological patients' outcomes will be collected, as well as the perception of quality of nursing care, and data on the system costs.
Conclusion: This research project is ongoing (approval from the Ethics Committee n.10/int/2018). Results will be useful to demonstrate the effects of a concrete experience of implementing Synergy Model.

NP-6

Immunoadsorption and plasmapheresis as treatment of the rejection after heart transplant

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The first heart transplant, at Bambino Gesù Pediatric Hospital, was carried out over 30 years ago; this practice still represents the gold standard for the treatment of terminal cardiomyopathies and complex congenital heart diseases, refractory to medical and/or surgical treatments.
However, important complications can arise, including those related to the development of rejection, either acute or chronic. The first episode generally occurs months after transplant, following an inflammatory response of T cells again the myocardium (cellular rejection), or because of the production of anti-HLA antibodies by the recipient's lymphocytes that identify the transplanted organ as a foreign body (antibody mediated rejection). Chronic rejection is characterized by a thickening of the wall of the coronary vessels', causing suffering of the new heart or a progressive fibrosis of the myocardium.
The first-rate treatment in our experience, aiming both at the prevention and treatment of rejection, consist in pharmacological therapy with steroids and immunosuppressive drugs. In the instance of this strategy's failure, previous experience in antibody mediated rejection, has promoted the use of monoclonal antibodies and apheresis, specifically plasmapheresis (a more aggressive method) and immunoadsorption.
These practices allow the selective elimination of cells or other harmful substances from blood, like immunoglobulins and immune complexes or inflammatory cytokines.
The care pathway for patients with graft rejection therefore provides a multidisciplinary approach.
In all steps of this process, starting with post-operative management, close collaboration between different professionals is needed. In this process nurses take care of:

- patient reception in the ward;
- management of the vascular access dedicated to the procedure;
- educating the patient and parents;
- psychological impact of rejection;
- monitoring of pre and post treatment blood indices;
- monitoring and recognition of any adverse reactions;
- prevention of care-associated infections in a compromised patient.

Overall, organ rejection prevention and treatment have significantly advanced using knowledge, skills and synergies of all the professionals involved in the care management of patients. But several and bigger studies are needed to improve the pediatric experience in heart transplant fields.

NP-7

Evaluation of pH and temperature in pediatric skin lesions. Prospective study

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The skin in newborns is characterized by peculiar features and particular attention in intensive care units is recommended.
We enrolled 20 newborns (14 males and 6 females) with pressure ulcers or skin epidermal stripping, from january 2017 to october 2017, at intensive care unit of FTGM Massa.
We recorded pH value and temperature value on 16 pressure ulcers and 11 epidermal stripping, comparing the results with value on intact skin.
Intact skin showed an average pH more acid than both epidermal stripping ($p<0.05$) and pressure ulcers. Temperature in pressure ulcers showed lower value than intact skin ($p<0.05$) and in epidermal stripping lesions showed higher value than intact skin.
The monitoring of pH and temperature may be useful in the evaluation of skin lesions in newborns. Further studies are need.

https://doi.org/10.1017/S1047951119000489 Published online by Cambridge University Press